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Contents

Frontispiece: Ludo van Bogaert.

ORIGINAL ARTICLES

ACUTE ENCEPHALITIDES. Prof. Ludo van Bogaert, M.D. .. 131
THE MORO REFLEX. Ross G. Mitchell, F.R.C.P.E. .. 135
PERCEPTION AND EYE MOVEMENTS. M. L. Johnson Abercrombie, PH.D .. 142
VARIATIONS IN PERCEPTUAL ABILITY AMONG TYPES OF CEREBRAL PALSY. K. Wedell, PH.D. .. 149
THE HYPOTONIC FORMS OF CEREBRAL PALSY. Ivan Lesny, M.D. .. 158
PITUITARY IMPLANTATION IN CEREBRAL PALSIED CHILDREN. I. Lesny, V. Vojya and V. Jelinek .. 167
HYDROCEPHALUS AND DISABILITY. K. M. Laurence, M.A., M.B. .. 170

VISITS

A VISIT TO PARIS. Leonard Haas, M.D., M.R.C.P. .. 180

EDITORIALS

The First Breath. T. Oppé .. 183
Right and Left. M. L. Johnson Abercrombie .. 187
Prognostic Value of the Neurological Examination of the Newborn. R. C. Mac Keith .. 189
Infantile Spasms. B. D. Bower .. 191
Akinetic Mutism. Peter H. Schurr .. 193
Second International Study Group on Child Neurology and Cerebral Palsy .. 196

OBITUARIES

Stella I. Albitreccia. Mrs. Judith M. Hillelson .. 198
Melanie Klein. D. Russell Davis .. 201

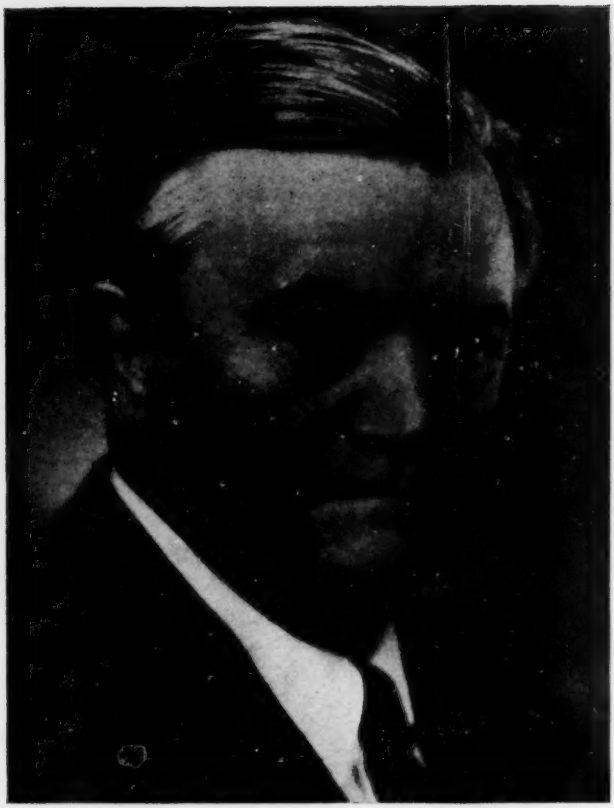
BOOK REVIEWS

Handbook of Speech Pathology. Edited by Lee Edward Travis. (T. T. S. Ingram) .. 203
The Chesty Child. Chest and Heart Association. (Christine Cooper) .. 204
Paediatric Neurosurgery. Edited by Ira J. Jackson and Raymond K. Thompson. (Peter H. Schurr) 206
Neurology of Infancy. By Anatole Dekaban. (T. T. S. Ingram) .. 207
Aids to Neurology. By E. A. Blake Pritchard. (John Burkinshaw) .. 208
Brain Memory Learning. By W. Ritchie Russell. (M. L. Johnson Abercrombie) .. 209
Maladies héréditaires du Métabolisme chez l'Enfant. By M. Lamy, P. Royer and J. Frézal. (Ross G. Mitchell) .. 210
New Opportunities for Deaf Children. By I. R. and A. W. G. Ewing. (L. F. W. Salmon) .. 211
Family Clinic. By *Sunday Times* Medical Panel, edited by Barbara Evans. (A. J. Dalzell-Ward) .. 211
Physics and Electronics in Physical Medicine. By A. Nightingale. (Philip Bauwens) .. 212

NOTICES

Vocational Guidance for the Handicapped. An International Seminar. M. R. Morgan .. 213
Films on Cerebral Palsy .. 214
A Hungarian Acta Paediatrica .. 214
International Journal of Neurology .. 214
World Commission on Cerebral Palsy. E. Clayton-Jones .. 215
A Job in Malaga? .. 234

PUBLICATIONS OF THE N.S.S. AND B.C.W.S. .. 216
ABSTRACTS .. 218
MAIN CONTENTS OF BACK NUMBERS .. 230



LUDO VAN BOGAERT

Dr. Ludo van Bogaert, a native of Antwerp, graduated in medicine at Brussels in 1922. He has spent the whole of his professional life in Antwerp and is Director of the Institut Bunge in that city. Dr. van Bogaert's researches and published writings cover many aspects of neurology, but he has paid special attention to the epilepsies, the encephalitides and the cerebral lipidoses. Apart from his own scientific work, he is known to his colleagues throughout the world for his genial labours as Secretary-General of the 1st International Congress of Neurological Sciences and as President of the World Federation of Neurology.

Acute Encephalitides

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This is the first of two review articles on the encephalitides. The second, on Subacute Encaphalitides, will be published in *Cerebral Palsy Bulletin* Vol. 2, No. 4.

IT is in infancy, and most often early infancy, that the most disastrous neurological diseases of adolescents and adults begin, often without our being aware of what is happening in the central nervous system, or at least without our recognising its gravity.

It is on the basis of the various forms of encephalitis and infantile encephalopathies that the substratum of the cerebral palsies, the epilepsies and numerous personality changes is constructed. The treatment of the sequelae can only be palliative and incomplete. It is from the moment of birth or even before, while the infantile cerebral disorders are waxing and waning, that the physician of tomorrow must initiate his treatment if it is to be effective. We are convinced that the prevention of many nervous and mental diseases encountered in later life must be based on the neurology of youth and infancy. So long as this is misunderstood by the public, and above all by paediatricians, most cases will not come to the neurologist until they have reached an irreversible stage.

The campaign against cerebral palsy must be waged by the obstetrician immediately before and after the birth of a child and during the first months of extra-uterine life.

I am discussing here the acute encephalitides seen in childhood in which early diagnosis is essential. To the clinician and pathologist the term 'encephalitis' means an inflammation of the brain accompanied by a clinical syndrome suggesting an affection of the cerebrum. In an appreciable number of cases the inflammation affects not only the cerebrum but also the meninges and spinal cord, so it is more correct to speak of 'meningo-encephalomyelitis'. For the sake of brevity, however, I shall here use the term encephalitis.

The diagnosis of encephalitis is made first on the history and secondly on the results of examination.

History

The onset of the encephalitis is sudden, with headache, fever, neck-stiffness and sometimes convulsions. Photophobia, nausea, agitation, somnolence, generalised trembling, shivering, and an inflamed throat have often been noticed by the parents beforehand. Every feverish child with these neurological signs should in fact be suspected of having an encephalitis. One should enquire if members of the family have suffered from any infectious diseases, since the encephalitides are transmittable. This can best be done by the general practitioner who first sees the child.

Some of these encephalitides are seasonal and related to certain climatic conditions. One must find out whether the child comes from a country where certain of these infections are endemic, such as Japan, California or Eastern Europe; whether he has lived in an area where there are mosquitoes or ticks; whether he has been in contact with people who breed birds (psittacosis), cattle, sheep or goats (Q fever); whether he has recently been drinking unboiled goat's milk (brucellosis and Q fever) or possibly contaminated water; and whether he has mixed with other children (poliomyelitis, mumps, exan-

thems). Above all one should ask whether he has recently been vaccinated against any disease.

Clinical Examination

If the child is gravely ill, our examination should be restricted to inspection, the colour of the skin, sweating, the way the child lies in bed, whether or not he can cough, whether his nasal secretions are excessive, and whether the pitch of his voice changes when he cries. One must look for evidence of mucus in the trachea, for abdominal swelling, and for signs of urinary retention or incontinence, and note the rhythm and symmetry of the respiratory movements.

This examination alone is often sufficient to establish whether the main lesion is supra- or infra-tentorial. Above all, the examination will decide whether the child needs tracheal intubation for the aspiration of mucus and whether he should be tube-fed or even given assisted respiration.

If the child's condition permits examination, one will look first at the ocular fundi. Ocular motor palsies suggest meningitis; the involvement of lower cranial nerves suggests poliomyelitis. In these cases Kernig's sign is positive. Pyramidal and extrapyramidal signs should arouse the suspicion of a post-exanthem encephalitis. Palpation of the muscles will give an indication of their tone and the extent of tenderness present (encephalomyelitis, poliomyelitis or polyneuritis). Equally important is evaluation of the tendon reflexes.

Laboratory Aid

The history and clinical examination will make it possible in a good number of cases to diagnose poliomyelitis, polyneuritis, or a postvaccinal or postexanthematous encephalitis. In many other cases one is left with the hypothesis that the encephalitis was caused by a virus or another infectious agent, and here one must seek the aid of the laboratory.

The information that the laboratory can provide will help to exclude a bacterial or spirochaetal infection, which can be treated with antibiotics and sulphonamides. In this respect lumbar puncture is particularly important. Examination of the blood, urine and cerebrospinal fluid gives useful information in a certain number of cases of rickettsial and viral infections; neutralisation and complement-fixation tests are most often used, but these necessitate serial blood-examinations or the use of time-consuming methods. These techniques are not very helpful in making an early diagnosis, in spite of the recent progress in this field.

The sick child has in fact often passed the acute phase of the disease before the laboratory results become available to the physician. This clearly does not mean that laboratory aid should not be sought in such cases. The results may well be of interest later in the course of the disease. It is important to know the exact cause of the disease, if only to diagnose analogous cases occurring in the same environment and at the same time. On the other hand, some viral diseases are liable to be accompanied by secondary infections which laboratory examinations can bring to light and which can be treated.

Nature of the Encephalitis

We must admit that laboratory investigations are not helpful in the early diagnosis of acute encephalitis. Is it suppurative or not? Is it a true encephalitis? Or is it an encephalitis caused by toxins arising from an extracerebral infectious process? All paediatricians agree that cerebral manifestations can be produced by an influenzal bronchopneumonia in predisposed children; they may be the most important feature during the course of an acute encephalitis. We also know that the cerebral symptoms may predominate and even provoke death through the electrolyte disturbances which one sees in certain infantile diarrhoeas. However, there are certain clinical signs which give us a clue if we give them due consideration.

In suppurative cases there is often shivering, leucocytosis with a shift in the Arneth count to the left, leucocytes with

toxic granules in the smears, and above all a focal infection (sinusitis, otitis, etc.).

In viral diseases the neurological syndrome is often preceded by a vegetative phase and a visceral phase (parotitis, hepatitis, adenopathies, pneumonia, cutaneous symptoms). There is also a relative leucopenia with lymphocytosis and often a diphasic temperature curve.

These minute differential signs are often useful, but this is true only in cases with focal signs that make it possible to establish the presence of an inflammatory lesion of the nervous system.

In some cases there is from the beginning a massive cerebral affection accompanied by coma, profuse sweating, convulsions, circulatory collapse, vomiting and possibly fever, with cardiac, renal and hepatic involvement. In this condition the affected organs are markedly oedematous, but it is difficult to tell whether the oedema is toxic or inflammatory in origin.

When this clinical picture is completed by a hemiplegia or double hemiplegia, one is dealing with a venous thrombosis or thrombophlebitis—in other words, the underlying cause of the encephalopathy is a haemorrhagic infarct of the cerebral tissue.

This encephalopathy, or serous or haemorrhagic encephalitis with hemiplegia, may be seen in a number of infections involving the head and face and in all the infectious diseases of childhood, even a simple gastro-enteritis. These are the commonest disorders and the most important ones in the pathogenesis of the sequelae which lead to 'cerebral palsy'.

Treatment in the Acute Stage

What can be done during the acute phase of the encephalitis or encephalopathy to mitigate the late consequences of the attack? In principle one should hospitalise and isolate the child and avoid useless and highly exhausting examinations, so as not to impair the child's resistance. Care must be taken of the skin, the bladder and the bowels, of the mouth and nasal cavities, the ears and the anus; one should not hesitate to administer sedatives, such as small doses of phenobarbitone, belladonna and tranquillizers.

Unfortunately there is no specific treatment for most viral encephalitides. It is useless to continue with the sulphonamides or antibiotics given in the early stages of the disease, once we are certain that we are not dealing with an infectious process or suppurative complications.

The administration of antibiotics or sulphonamides will be determined not only by the type of antibiotic but also by such factors as the child's state of consciousness, his reaction to these drugs and his renal function. On the other hand, one cannot over-emphasise the importance of watching the patient's nutrition. This is particularly true if the child is either incapable of taking nourishment (bulbar lesion) or is vomiting. When the child is lethargic it may be necessary to give fluids and dextrose parenterally in sufficient quantities for two or three days. In stuporous patients it is preferable to give feeds by gastric tube, so that it is possible to add salt, milk, eggs, fruit juice, vitamins and sugar. In serious cases complicated by anaemia, blood-transfusion should be considered.

One should pay close attention to fluid balance, for these children become dehydrated as a result of diarrhoea, vomiting, sweating, fever and hyperventilation, and this often happens rapidly.

Symptoms of cerebral oedema are the only contraindication to rehydration by mouth or parenterally. The fluid balance is seldom upset in cases of encephalitis except when there is excessive vomiting. On the other hand, there is often a serious disturbance of electrolytes in the toxic encephalopathies. Here the hypodermic or intravenous administration of isotonic saline in proportion to body-weight should be considered immediately and the restoration of ionic equilibrium must be carefully watched.

So far as the treatment of cerebral symptoms is concerned, one should pay particular attention to convulsions, for which chloral is the best drug. If necessary

this should be given intramuscularly every 20 minutes, followed by phenobarbitone. If it proves impossible to check the convulsions, ether anaesthesia with some oxygen may be given. Finally, in status epilepticus one may be forced to administer chloral intravenously in an isotonic solution of salt and sugar, or by drip. Small doses are advisable because chloral depresses the respiratory centre. At the same time one should try to control the temperature and fluid balance. Once the status epilepticus has subsided and the child is having only a few convulsions one may return to amylobarbitone or phenytoin.

It is obvious that certain dysfunctions arising in the course of encephalitis or the encephalopathies may themselves produce cerebral damage. This makes the prognosis of the disease worse.

In a prolonged state of unconsciousness one should use sedatives with caution and ensure adequate oxygenation, nutrition and hydration. Particular attention should be given to the bladder and bowels. Many pulmonary complications can be avoided by the timely aspiration of mucus from the trachea or by intermittent postural drainage. The child's position should be changed often. Tracheotomy or artificial respiration must be considered in any case where there is serious impairment of the respiratory centres.

In some viral infections treatment has an almost specific effect. The general principles of the indications, administration and dosage of antibiotics and sulphonamides in suppurative disorders are described in every modern textbook and need not be mentioned here.

SUMMARY

Encephalitis still remains a ticklish and important problem for the practitioner because the prognosis and treatment of these cases are of prime importance. A broad outline is here given of present views on the differential diagnosis, laboratory examinations and therapy.

RÉSUMÉ

Les encephalites aiguës

L'encéphalite demeure encore un problème délicat et de grand intérêt pour le praticien car le pronostic et le traitement de ces cas sont de première importance. L'auteur esquisse à grands traits l'état des connaissances actuelles sur le diagnostic différentiel, les analyses de laboratoires et la thérapeutique contemporaine.

ZUSAMMENFASSUNG

Akute Encephalitides

Die akute Encephalitis bleibt noch ein heikeles und bedeutentendes Problem für den praktischen Arzt, denn die Prognose und die Behandlung dieser Fälle sind von höchster Wichtigkeit. Der Verfasser entwirft einen weiten Umriss von dem, was man heute von der Differentialdiagnose, den Laboratoriumsanalysen und der gegenwärtigen Therapie kennt.

The Moro Reflex

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ON May 7, 1918, at a meeting of the Natural History and Medical Society in Heidelberg, Prof. Ernst Moro gave an address on the first three months of life. At the end of his paper, almost as an afterthought, he mentioned an observation he had made—that if the pillow on either side of a young infant is struck with the hands, a peculiar movement results, in which the arms move out symmetrically and then come together in an arc. His address, which was published later in the *Münchener Medizinische Wochenschrift*, attracted much interest, and the movement described soon became known as the Moro reflex. In the ensuing years, many papers were written on the diagnostic and prognostic value of the reflex in clinical work and on the various methods of eliciting the response. More recently, especially since the introduction of high-speed cinematography, attention has been directed towards analysis of the various components of the reflex and their physiological meaning.

Fig. 1. The Moro reflex, showing the primary response of extension.

Description of the Movements

The Moro reflex is a composite series of movements, not all of which may be observed in the individual infant. Moreover, the nature of the movement pattern gradually alters with increasing maturity during the first few months of life. It is not possible, therefore, to give a single description of the Moro reflex which will be applicable at all ages and to all infants. The account which follows refers to the reflex as seen in its entirety in the infant of a few days old.

The initial part of the response is extension and abduction of the upper extremities, with extension of the spine and retraction of the head (Fig. 1). The forearms are supinated and the digits tend to extend and fan out, with the exception of the distal phalanges of the index finger and thumb, which may be flexed in the shape of a 'C' (McGraw 1937). Sometimes there is slight tremor or even rhythmic shaking movements of the limbs. After a momentary interval, almost imperceptible clinically but revealed by high-speed photography (Goldstein *et al.* 1938), the upper extremities describe an arc-like movement, bringing

the hands towards one another in front of the body, and finally return to the position of flexion and adduction. The movements in the lower extremities are less pronounced. Usually both extend and abduct with the upper extremities, although there may be a slight movement of flexion first. If the lower extremities are extended when the stimulus is applied, however, flexion movements may be more prominent.

Several attempts have been made to grade or classify the movements of the Moro reflex. McGraw (1937) described three phases during maturation—the initial or newborn phase, the second or transitional phase, and the final or mature phase, when all gross movement seems to be suppressed and the reaction consists of a fine body jerk associated with blinking. She observed that in the newborn phase the movement response is usually followed by intense crying.

Gordon (1929) recognised three degrees of intensity of the movements and designated them group 1+ (shrugging of shoulders and slight forward movement of arms); group 2+ (arms approach one another, elbows bent and fingers stretched); and group 3+ (arms describe a complete arc, with slight tremor of hands and stretching of legs). Stirnimann (1943) classified the Moro reflex according to the type of response and found the incidence of the various types to be as follows:

Equally strong abduction and adduction of extremities ..	2%
Preponderant abduction ..	60%
Preponderant adduction ..	23.5%
Asymmetrical reaction ..	7.5%
Absent or indistinct reaction ..	6%
General convulsive movements	1%

De Grignon (1955) preferred to consider the movements of the upper and lower extremities as two separate reflexes—'Moro supérieur' and 'Moro inférieur'. In this connection it is worth noting that Stirnimann recorded movements of the lower extremities in only 82 per cent of cases showing movements of the upper extremities.

Methods of eliciting the Reflex

A sudden jolting movement, such as that produced by striking the table on either side of the infant, will elicit an immediate response; this was the method advocated by Moro and practised by many subsequent workers. The table must be so constructed that a change of position is produced by the blow, for according to Sanford (1933) it is impossible to obtain a satisfactory response if the support is too solid. Freudenberg (1921) showed that many other stimuli, such as the application of heat or cold, a tap on the abdomen, and even hiccoughing could evoke a response. He also noted that the reflex is not elicited by ventral flexion of the head with the body in the supine position, but may appear if the child is lifted without supporting its head. Sanford (1933) reported a satisfactory response obtained by holding the feet and head firmly in a horizontal position and allowing the head and upper part of the body to drop a few inches—i.e., allowing extension of the head on the body. These various authors' observations have been amplified and elaborated by André-Thomas and his colleagues (André-Thomas and Hanon 1948), who noted that, when the body is sharply displaced backwards, there is no response if the head is held in flexion in constant relationship to the body, but the reflex is elicited if the head is allowed to go free or is brusquely extended. They concluded that the essential stimulus is the sudden extension of the head relative to the trunk.

The elicitation of the Moro reflex by loud noises has been variously interpreted. Freudenberg (1921) stated that aural stimuli would not produce the reflex. Many subsequent workers, however, have successfully used a loud noise as the stimulus. Ford (1952) stated that 'Sudden loud noises produce auditory reflexes in normal infants shown by . . . the Moro reflex. Thus the presence or absence of hearing may be determined even in the newborn'. Others have considered that such responses to loud noises are the result of vibration rather than auditory in nature. It seems

likely that both methanisms may operate in different situations, although at best noise must be regarded as a poor method of eliciting the Moro reflex.

As McGraw (1937) has pointed out, the list of effective methods of eliciting the reflex suggests that it is the quality of suddenness in the stimulus which is important. André-Thomas and his colleagues, however, believe that the essential stimulus is sudden extension of the head relative to the trunk, and that all the methods of elicitation described operate in this way (Saint Anne-Dargassies 1954). Such a mechanism seems improbable when certain stimuli—e.g., a tap on the abdomen or a loud noise—are employed, but the possibility that they produce an initial minimal extension of the neck, perhaps by inducing a startle response, cannot be denied (the meaning of 'startle' is discussed below).

Disappearance of the Reflex

The Moro reflex is strongest during the first eight weeks or so of life; thereafter as each week passes it becomes increasingly difficult to elicit and the movements become progressively less gross. Moro himself noted that after the first three months the reflex was only obtained with difficulty but that it might persist longer in premature infants. Gordon (1929) found that the reflex disappeared at the end of the fourth month; thus only 13 of 63 infants failed to respond in the third month, as against 50 of 63 in the fourth month. From observations on 300 normal infants, however, he noted that the response might occasionally be observed during the fifth month. McGraw (1937) preferred to regard the Moro reflex as maturing rather than disappearing. She found that most infants change at about 90 days from the newborn phase to the transitional phase and at about 130 days to the final 'body-jerk' phase, which then becomes the characteristic response. De Grignon (1955) also considered that the Moro reflex persists longer than generally supposed, modifying itself slowly and becoming an integral part of future activity.

Clinical Significance of the Moro Reflex

Much of the early literature on the Moro reflex was concerned with its clinical use as a diagnostic aid, and can be summed up briefly under three headings:

1. *Absence of the reflex* in the newborn infant often indicates cerebral damage. The intensity of the response during the first few weeks may be greatly diminished by birth injury to the brain or by general muscular weakness from any cause. With improvement in the clinical condition the intensity of the reflex may increase. Sanford (1933) believed that the Moro reflex could be used to distinguish among babies with signs of cerebral damage those who had cerebral haemorrhage from those who had not. He reported that in cerebral haemorrhage the reflex was present on the first day, then disappeared and only returned slowly after the sixth day. In cases without haemorrhage (which he called cases of cerebral oedema) the reflex was absent on the first day but gradually returned during the following four days.

2. *Asymmetry of the response* may be a sign of local injury. Thus damage to the brachial plexus or fracture of the clavicle may inhibit the reflex movements on the side affected. It should be noted, however, that Stirnimann (1943) found an asymmetrical response in 7.5 per cent of normal infants.

3. *Persistence of the reflex* after the age of six months is an indication of neurological abnormality, and is common in children with mental retardation or with disorders affecting the cerebral control of motor function. Thus in children with severe cerebral palsy, the Moro reflex may persist in partial or complete form for years. Caution should be used in interpreting persistence of the reflex in premature infants, for, as recognised by Moro himself, the response may be elicited for an unusually long period in such infants, the

duration corresponding to the degree of immaturity.

Physiological Meaning

In his original paper, Moro suggested that the reflex he described was a primitive movement of clasping or embracing (Umklammerungsreflex) analogous to that by which the young of such creatures as the ape and the bat instinctively cling to their mothers (Fig. 2). Thus from the beginning the Moro reflex was regarded as an embrace reflex, and as late as 1937 no other outstanding theories as to its origin and purpose had been proposed (McGraw 1937). Although Moro stressed the clasping nature of the movements, however, careful analyses of the reflex pattern in recent years have shown clearly that the movement of extension is the essential part of the Moro reflex (Fig. 1), and that the approximation of the upper extremities is a secondary component. It has been suggested that the latter is simply a returning of the limbs from the position of extension and abduction to the normal position of flexion and adduction, but the movement is too positive to be regarded merely as a passive return to rest. It is known, however, that on cessation of a stimulus, a reflex very frequently exhibits a rebound contraction (Sherrington 1913) and this could explain the active nature of the flexor component.

Goldstein and others (1938) suggested that the extension response of the Moro reflex might be similar to the kind of muscle pattern seen in decerebrate rigidity, especially as both are responses which appear if the regulatory cortical influence is less than normal. However, in decerebrate rigidity in man, the upper limbs are usually flexed and adducted, rather than extended, a characteristic which has been attributed to the lack of locomotor function of the upper limbs in man (Walshe 1919). It is true that rigid extension of the upper limbs in both adults and children has been described as decerebrate rigidity (Wilson 1920) but in such cases, as in decerebrate rigidity in monkeys and lower animals, the upper limbs are strongly adducted at the shoulders, with hyperpronation of the forearms and flexion of the digits—in complete contrast to the abducted arms, supinated forearms and extended digits of the Moro reflex. Neither in human beings nor in experimental animals, therefore, is there any position of decerebrate rigidity corresponding to the position observed in the Moro response.

Wieser and Domanowsky (1959) considered that there was no good evidence to support either the embrace or the decerebrate rigidity hypothesis, and regarded the Moro reflex as a complex response resulting from the interplay of several postural reflexes.

Fig. 2. Drawing from Moro's original paper, illustrating his concept of the reflex as a primitive movement of clasping. (Reproduced with the publishers' kind permission from *Münchener Medizinische Wochenschrift*, 1918, **65**, 1147).

Whatever the explanation of the Moro reflex, it is plain that it must be regarded as a response of immaturity. McGraw (1937) suggested that maturation of the response takes place because myelinization progresses, neural pathways become linked, and cortical control increasingly restricts the somatic response.

Relationship to Other Reflexes

Some earlier workers, such as Peiper and Isbert (1927), considered that the Moro reflex was a labyrinthine reflex. This view was challenged by Freudenberg (1921) and Gordon (1929) who, however, appear to have believed that Peiper and Isbert were referring to the tonic labyrinthine reflexes, whereas in fact they meant that the Moro was a stato-kinetic reflex. The Moro reflex is not, of course, a tonic labyrinthine reflex since it does not depend on the position of the head in space. It may in part be a response to stimulation of the labyrinth by sudden acceleration—i.e., a stato-kinetic reflex—but it cannot be attributed entirely to this, since, according to André-Thomas, sudden alteration of the position of the head in space without altering the relationship between the head and trunk does not evoke the reflex. It seems most probable that the Moro reflex results from summation of a number of different reflexes originating in the neck muscles and in the labyrinths.

The Moro reflex has at various times been described as the 'embrace', 'clasp', 'alarm', 'fear' and 'startle' reflex; in fact none of these epithets can properly be applied to it. 'Embrace' and 'clasp' were the original and still widely used terms, but, as has been pointed out, their use is based on a misconception of the nature of the Moro reflex, which is essentially a movement of extension and not of flexion. The reflex should probably not be regarded as a reaction to 'alarm' or 'fear', because emotional responses cannot be clearly differentiated by observation of the newborn infant, and their interpretation in terms of adult emotions is inappropriate.

It is perhaps the word 'startle' that has caused most difficulty, and in fact this term was, and in many textbooks still is, employed as a synonym for the word 'Moro'. Confusion has arisen because the word 'startle' can be used in different ways, either in a general non-specific sense to convey the meaning of surprise or as a precise descriptive term for a well-defined reflex. This was described in detail by Strauss (1929) as the response of the adult human to a sudden stimulus, and consists of forward movement of the head; closing of the eyes and distortion of the features; raising and drawing forward of the shoulders; abduction, forward elevation and inward rotation of the upper arms; flexion of the elbows; pronation of the forearms; clasping of the hands; contraction of the abdomen; forward movement of the trunk; and flexion of the lower extremities at hips and knees. Thus the movement pattern differs considerably from the Moro reflex, being primarily a response of flexion and not of extension. This 'startle pattern' has been recognised as occurring in the young infant and as being quite distinct from the Moro reflex (Hunt and Landis 1938, Wieser and Domanowsky 1957). It is not surprising that the difference is difficult to discern clinically, for the startle pattern is often not fully developed before six weeks of age, and the response is so rapid that it ordinarily escapes the human eye. Earlier workers seem to have recognised its existence, however, and Gordon (1929) discussed its relationship to the Moro reflex, although he used the term 'fear reflex' instead of 'startle pattern'. Others observed the gradual strengthening of the startle pattern concurrently with the diminution in intensity of the Moro reflex, and interpreted the former as the mature stage of the Moro response. Wieser and Domanowsky (1959) appear to regard the two responses as phases of a single 'alarm reaction' (Schreckverhalten).

It seems clear from the work of Hunt and Landis and their colleagues with ultrarapid photographic methods that the startle and Moro reflexes are quite separate and different, that both may be elicited in the

same infant by the same stimulus, and that the rapid startle pattern usually precedes the slower Moro reflex, but may be obscured by it (Goldstein *et al.* 1938). In view of these differences, it seems advisable to cease using the word 'startle' in a general sense for any response to a sudden unexpected stimulus, and to confine its use to the specific 'startle pattern' of Hunt and Landis.

Other descriptive terms have been suggested for the Moro reflex. McGraw (1937) described it as a 'defensive' or 'protective' reaction. Stirnimann (1943) thought that the designation 'concussion reaction' was more appropriate. On the whole, however, it seems preferable to retain the name 'Moro reflex', which has the merits of being short, precise and unlikely to be misinterpreted.

SUMMARY

The Moro reflex is a composite series of rapid movements made by young infants in response to sudden stimuli. In the response of the Moro reflex, the essential component is the movement of extension; in which the Moro response contrasts with the 'startle' response, which is a very rapid movement predominantly of flexion. This startle pattern may sometimes precede or may be obscured by the Moro response.

The Moro reflex becomes progressively more difficult to elicit from the third month onwards and by the fifth month has usually disappeared, at least in recognisable form. The reflex is of some value in clinical diagnosis and its persistence beyond the normal period is an indication of neurological abnormality. It should not be called an 'embrace' or 'clasp' reflex, and the word 'startle' should be reserved specifically for the 'startle pattern'.

RÉSUMÉ

Le réflexe de Moro est composé d'une série de mouvements rapides effectués par les jeunes nourrissons en réponse à une stimulation brutale. La composante essentielle est le mouvement d'extension. En ceci la réaction de Moro diffère de la réaction de sursaut qui est un mouvement rapide, surtout en flexion. La réaction de type arrêt peut précéder ou être masquée par le réflexe de Moro. Celui-ci devient de plus en plus difficile à être mis en évidence à partir du 3ème mois; vers le 5ème mois il a en général disparu ou tout au moins il n'est plus reconnaissable. Ce reflexe a une certaine valeur de diagnostic, et sa persistance au delà de la période normale indique une anomalie neurologique. Il ne faut pas l'appeler reflexe d'embrassement ou d'étreinte et le terme de sursaut devrait être réservé à la réponse qui lui correspond.

ZUSAMMENFASSUNG

Der Morosche Reflex setzt sich aus einer Reihe schneller Bewegungen, die die jungen Säuglinge als Antwort auf einen plötzlichen Anreiz ausführen, zusammen. Sein wesentlicher Bestandteil ist eine Streckbewegung. Dadurch unterscheidet sich die Morosche Reaktion von der 'Auffahrreaktion', die eine sehr schnelle, hauptsächlich durch Beugen gekennzeichnete Bewegung ist. Eine solche 'Auffahrreaktion' kann manchmal dem Moroschen Reflex vorangehen oder von ihm verdeckt werden.

Von 3 Monat ab wird es immer schwieriger, den Moroschen Reflex auszulösen und im 5 Monat ungefähr verschwindet er oder wird wenigstens unerkänntlich. Dieser Reflex besitzt einen gewissen diagnostischen Wert und sein Fortbestehen über die normale Periode hinaus zeigt eine neurologische Anomalie an. Man darf ihn nicht 'Umarmungs-' oder 'Umklammerungsreflex' nennen und man müsste den Ausdruck 'Auffahren' eigens für die 'Auffahrreaktion' aufbewahren.

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Perception and Eye Movements: Some speculations on disorders in cerebral palsy*

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THE disorders of visual space perception observed in some children suffering from cerebral palsy have received a great deal of attention in the last few years. There has, however, been little systematic work on the relation between perceptual disorders and eye defects which are common in children with cerebral palsy. Some workers with cerebral palsied children have adumbrated the possible connection of ocular and perceptual disorders (Dunsdon 1952, Schonell 1958, Guibor 1953), but it is probably fair to say that in general the assumption is made, either explicitly (e.g. Woods 1957) or implicitly (if one may judge from the design of investigations) that the perceptual disorders are of central origin, and that any peripheral disorders that there may be do not contribute significantly to the perceptual disorder.

This assumption—that the difficulty is one of manipulating symbols rather than of defect in sensory input (however determined)—receives support from the cases that have been cited of children with space perception difficulties who are said to have no physical defect of vision (e.g. Floyer 1955). Most studies of perception, however, fail to give any report of the condition of the eyes, and though it is probable that gross defects were absent or were allowed for, it is possible that less obvious disorders were present and relevant. Even in the report of the ambitious project of Cruickshank, Bice and Wallen (1957), in which the figure-background perception of 211 spastic, 114 athetoid and 110 non-handicapped children was investigated, no ophthalmological data are given: the possibility that the subjects had any sensory or peripheral anomalies is dismissed with the statement that, because all the children have received much medical attention, it was likely that any correctible impairments had been corrected and no cases were used where the medical record indicated any uncorrected defect.

The work of Wood (1955) and Wedell (1959) is exceptional. In comparing the performance in perceptual tests of right and left hemiplegics Wood took note of certain visual disorders within the two groups but seems not to have compared the performance of those with and those without good vision. Wedell's work on perception in cerebral palsy included a deliberate attempt to investigate the relationship of visual handicap to performance in perceptual tests, but he found no significant relationship. It is possible, however, that the different eye disorders were not sufficiently specified, and that the heterogenous nature of the visual handicaps may have obscured any relationship there might have been.

Several different kinds of eye disorder occur more frequently in cerebral palsied

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than in normal children, such as errors of refraction, visual field defects, and various developmental anomalies—e.g., retrolental fibroplasia (see Illingworth 1958 for a useful summary). Here I am concerned only with disorders of movement of the eyeball.

Eye Movement in Cerebral Palsy

The outstanding manifestation of cerebral palsy is disorder of movement, and this certainly extends in many cases to disorder of movement of the eyes, both of vergence (when the two eyes move in opposite directions) and of version (when they move in the same direction). Estimates of the incidence in cerebral palsied children of disorders of *vergence* (including squint) vary between a quarter and three-quarters, depending on the population studied and on the degree of disorder recognised (Asher and Schonell 1950, Breakey 1955, Floyer 1955, Guibor 1953, Schachat *et al.* 1957, Wolfe 1950, Woods 1957). Disorders of *version* movements are also reported—nystagmus, difficulties of fixation, and paralysis of vertical movements. It is reasonable to suppose that still other version movements may be affected in some cases—e.g., the scanning movements used in exploring a visual pattern, the movements used in reading when the eyes fixate at intervals along the line of print, and even possibly the smaller and larger oscillations of the globe that occur during fixation of an image and that are important in maintaining the visual acuity of normal people (Adler 1959, Ditchburn *et al.* 1959).

There seems to have been very little attempt to study the possible effect on perception in cerebral palsy of these eye movement disorders, but work in two other fields is relevant to this subject—namely, the disorders of space perception of adults with brain damage, and the specific* reading difficulties of some normal people.

Brain Damage and Reading Difficulties

In cases of brain damage, disorders of eye movements have frequently been reported (Critchley 1953), and there has been considerable debate as to the role played by them, whether they cause, or are caused by, the perceptual defect: it is obvious that if a person does not understand what he is looking at his eye-movements are likely to be more erratic than those of one who knows where to look for what. On the other hand, if the eye movements are unco-ordinated, jerky or slow, and the fixation periods too long or too short, the sequence of images received on the retina will be abnormal and the difficulties of integrating them into meaningful perception may be increased. In one case at least there is good evidence of a causal relationship between disordered eye movements and the breakdown of a perceptual skill. Warrington and Zangwill (1957), reporting a case of 'pure' dyslexia in a brain-damaged adult, concluded that in this patient the difficulty in reading was caused by anomalies of eye movements following on homonymous hemianopia. The patient's reading improved when he was given systematic practice in making lateral eye and head movements along the line of print while reading. The authors were careful to differentiate this case from other cases of dyslexia in which defect of recognition cannot be ruled out.

Disorders of vergence movements have been reported in some cases of brain damage and may have occurred in other cases, where double vision and loss of stereoscopic vision have been noted (Critchley 1953). The importance of their contribution to the disordered perception of the patients is conjectural.

As to the disordered version movements which are known to occur in cases of reading difficulty in normal people, again it is debatable which is cause and which effect. Vernon (1957), discussing backwardness in reading, says: 'It has been made abundantly clear that, except possibly in cases of ocular defect, it is failure to attend to and to comprehend what is read that

* I.e., reading difficulties occurring in the absence of factors which are correlated with them in other cases, such as low intelligence or lack of formal schooling.

causes irregular eye movements and regressions.'

On the other hand, there is evidence that disorders of vergence movements cause reading difficulties in some subjects (Eames 1953, Robinson and Huelsman 1953, Smith 1954, Vernon 1957, Woolf and Woolf 1957), and among brain-damaged children Eames (1959) found more muscle imbalance in the poor readers than in the good readers. The case for central versus peripheral causal factors is still accepted, however, by some people—e.g., by Goldberg (1959), who writes 'Defective vision and muscle unbalance do not have any significant role in the aetiology of a condition influenced by spatial confusion and poor visual memory.'

Version Movements and Perception in Cerebral Palsy

Even if the evidence from brain-damaged adults and backward readers for the causal relationship of eye movement disorders and perceptual disorders has been clearly established in only a few cases, there is reason to look a little more carefully into the conditions in cerebral palsy. The human eye muscles are richly supplied with nerves —1 nerve fibre to 5 or 10 muscle fibres, compared with 1 to 120 in the soleus muscle (Adler 1959)—and it is difficult to believe they are there for any other reason than to subserve perception in some way or other.

I am not suggesting that all perceptual difficulties of cerebral palsied children are due to inco-ordinated movements of the eyes, for clearly the perceptual disorders of cerebral palsied children do not form a homogenous class, any more than do those of brain-damaged adults or of backward readers (see, for instance, Caldwell 1956, Cruickshank *et al.* 1957, Dunsdon 1952, Floyer 1955, Holden 1952, Strauss and Kephart 1955, Taylor 1959, Williams 1958). Some cerebral palsied children, compared with normal children of the same age and general intelligence, have special difficulty with tests involving the abilities to match or memorise shapes, to interpret pictures, or to distinguish a figure from background. These seem to be specific disabilities of perception, in the sense that they would not be expected from consideration of the child's behaviour in other matters and are not necessarily related to the degree of handicap of manual dexterity.

Difficulties in tasks involving hand-eye co-ordination also occur in some cerebral palsied children. These are indicated in difficulties of dressing, or in constructive tests, such as assembling a manikin or building with bricks, tests concerned more obviously with manipulation in three-dimensional space and possibly with the orientation of the body in space. They resemble the conditions in some brain-injured adults (see, for instance, McFie, Piercy and Zangwill 1950, Critchley 1953). I am not going to argue here that eye movement disorders play a major part in such bizarre conditions, but the ability to discriminate shapes or patterns is basic to visuo-motor skills, and it is in the learning of pattern discrimination that eye-movements may be important.

It seems that, compared with normal children, some cerebral palsied children are prone to be *backward* in learning perceptual skills (see Cruickshank *et al.* 1957), and the speculation made here is that their slowness may be due in part to inefficient co-ordination of the movements of the eyes going back to early infancy. This speculation, which is in line with Piaget's (1948) views on perceptual development, derives directly from Hebb's (1949) argument that eye movements 'contribute, constantly and essentially, to perceptual integration even though they are not the whole origin of it'. 'I propose,' Hebb says, 'that the human capacity for recognising patterns without eye movement is possible only as the result of an intensive and prolonged visual training that goes on from the moment of birth during every moment that the eyes are open, with an increase in skill over a period of 12 to 16 years at least.'

The delayed learning of perceptual skills by some cerebral palsied children would then fall into line with their delayed develop-

ment of other motor skills, of sitting, standing or walking. If this is so, it might be possible to improve the effectiveness of the eye movements of babies in the population at risk, perhaps by letting the eyes follow moving lights, or by offering specially attractive targets such as those investigated by Berlyne (1958).

Vergence Movements and Perception in Cerebral Palsy

I shall extend Hebb's argument which was concerned mainly with fixation, scanning or outlining movements, to vergence movements and shall speculate how inefficiencies of these may slow down learning to discriminate shapes. The most obvious defect of co-ordination of vergence movements is squint, in which the axes of the two eyes can be seen to deviate so that they are not directed simultaneously to the same object. A less obvious defect of vergence movements is heterophoria (latent squint or latent deviation), in which although the eyes usually appear straight they have a tendency to deviate. This can be detected by covering one eye momentarily; when the cover is removed, the eye will be seen to have deviated from the fixing eye, and to come back more or less slowly. In heterophoria the eye movements are not always co-ordinated in such a way that when one eye fixates an object, the other automatically fixates it, as happens in perfect binocular functioning. If the extent of deviation is great, and recovery movements are weak or slow, the condition will affect perception in a way similar to squint, though to a lesser degree, and intermittently.

In squint and heterophoria a double image may be perceived. This may be confusing, for an object may appear to be in two places at once if the images are separated, or if the images are closer together and overlap the outline of the object will appear to be more complicated —a diamond may be seen as having more than four corners. If the images are brought still nearer together there may be a blurring of the outline, and inadequate definition of the contours. Moreover, with changing degrees of co-ordination of the two eyes, the extent of fusion or separation of the images may fluctuate from moment to moment. It is suggested that this indefiniteness and inconstancy of perception not only makes it more difficult to discriminate shapes at any single presentation but also makes it more difficult to learn to do so.

The discomfort caused by seeing double may be reduced by suppressing one of the images. If suppression occurs persistently in the same eye, ultimately its visual acuity deteriorates and the person becomes virtually permanently monocular. In this case stereopsis (depth perception), which depends on the fusion of two slightly dissimilar images on the two retinas, cannot of course be attained, and the person has to rely for depth perception on monocular clues such as overlap, perspective and shading (Adler 1959, Gibson 1950, Vernon 1952). But there may be *alternating* suppression of the images of the two eyes. The difficulties that arise through using the eyes alternately can be imagined by holding a finger so close to your nose that your eyes do not converge sufficiently upon it, and two fingers are seen: if first one eye is closed and then the other, the finger seems to jump from side to side. Alternating suppression may slow down reading because different parts of the line, or different lines, may be alternately fixated: the words get jumbled and some may be omitted and some read twice. Many of the squints of cerebral palsied children are of the alternating kind, with alternating suppression and good visual acuity preserved in both eyes (Adler 1959).

The suggestion is made then, that inefficiencies of vergence movements may contribute to disorders of perception in cerebral palsy and this in two ways. First, one may expect that double vision and alternating suppression may result in a confused or inconstant pattern being perceived, an object appearing to be in two places at once, or to have a confused contour, or to be in a different place from one moment to the next, so that learning to

discriminate patterns may be slowed down. Secondly, without good fusion of two images binocular stereoscopic vision will be absent or inconstant.

Value of Stereoscopic Vision

Now in some otherwise normal people stereoscopic vision fails to develop, even if the images are fused, and this seems not to be regarded as a very great handicap because in ordinary circumstances sufficient monocular clues may be available to appreciate depth. However, I want to suggest that binocular stereoscopic vision may be important in other ways than in the well-accepted one of providing more information about the relations of objects in space. It helps to differentiate one part of the visual environment from another, for the object of binocular regard, when its two images are fused stereoscopically, becomes clearly defined and almost palpably differentiated from the rest of the panorama. If you wake up in a strange room with one eye still on the pillow, the flat scene is often incomprehensible until you lift your head. Seeing two sides of a thing makes it more real, more understandable, and distinguishes it more sharply from the background.

The assistance that binocular stereoscopic vision gives in this way to differentiating figure from background may be important also in interpreting pictures, especially those which do not give sufficient monocular clues to space perception, such as overlap, perspective and shading. The initial difficulties of seeing 'meaning' in such pictures can be illustrated with the 'Hidden Man' (Fig. 1). At first sight this looks just a muddle of black and white patches, but when you see the man's face it seems to move forwards, to acquire not only shape but body, solidity or roundness, and to leave the background behind. This perceiving of depth in a flat picture is comparable with the impression of depth one gets when stereoscopically different images are presented to each eye. The experience of depth in this latter case results from the perceiving as a single image two images which are sufficiently dissimilar to fall in part at least on disparate regions of the retinas (Ogle 1950). It is tempting to wonder whether perhaps the projecting of depth into a single flat picture may be achieved by a variation of this method—namely by deviating the eyes sufficiently to get the two (similar) images on disparate parts of the retinas. But whatever the mechanism is by which we project depth into a flat picture, it seems to me likely that it depends on having previously learnt to perceive depth binocularly in real objects.

I am suggesting then that the difficulties

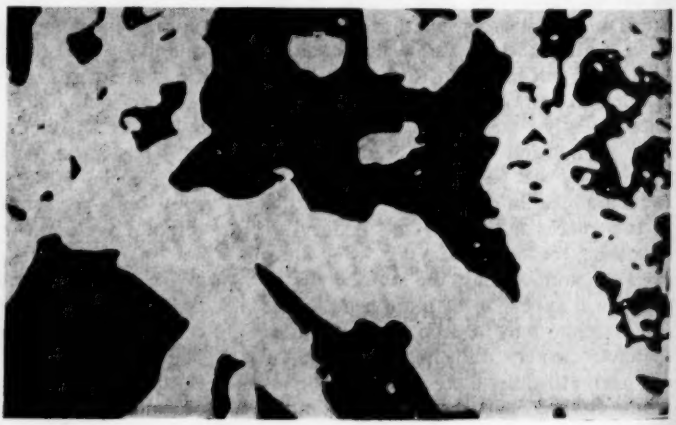

Fig. 1.
'The Hidden Man.'
(Reproduced by kind permission of the Editor from the *American Journal of Psychology*, 1954, **67**, 550.)
(*If after you have looked for some time you do not see the man, turn to page* 215.)

some cerebral palsied children have of interpreting pictures, and of distinguishing figure from background, may be related to inefficiencies of binocular stereoscopic vision. Their world may be flatter, less easily differentiated and endowed with meaningful structure than ours, both their real world and the pictured worlds. Even in normal people the attainment of full binocular vision with depth perception develops slowly, and does not become complete until the sixth year or later (Mann 1940). It is likely that in some cerebral palsied children full binocular vision, dependent as it is on motor co-ordination, develops still more slowly than in normal children, and in those with severe defects of vergence movements, not at all.

SUMMARY

The aim of this paper is to draw the attention of psychologists to the possible relevance of disordered eye movements to disordered perception in cerebral palsy, and to draw the attention of students of eye movements to the possible interest of cerebral palsy in providing natural experiments for the study of the effects of abnormalities of motor and sensory experience on the development of perceptual skills. It is hoped that if a causal relationship can be established, and if means can be found to treat the oculomotor disorders in early life, it may be possible to prevent the development of some of the disorders of perception.

RÉSUMÉ

Le but de cet exposé est d'attirer l'attention des psychologues sur le rapport pouvant exister entre les troubles des mouvements oculaires et de la perception dans l'infirmité motrice cérébrale; il s'agit, d'autre part, d'attirer l'attention de ceux qui étudient les mouvements oculaires sur l'intérêt éventuel de l'infirmité motrice cérébrale, comme champ d'expérience naturel pour l'étude des effets des anomalies motrices et sensorielles sur le développement de la perception. On espère que, si un rapport de cause à effet peut être établi et si on réussit à trouver le moyen de traiter précocement les troubles oculo-moteurs, on devrait pouvoir prévenir l'installation des certains troubles de la perception.

ZUSAMMENFASSUNG

Diese Arbeit zielt darauf hin, die Aufmerksamkeit der Psychologen auf die Möglichkeit eines Zusammenhanges zwischen den Störungen der Augenbewegungen und des Wahrnehmungsvermögens bei Zerebrallähmung zu lenken.

Anderseits ist für diejenigen, die die Augenbewegungen studieren, Zerebrallähmung vielleicht interessant als natürliches Experimentgebiet für das Studium der Wirkung motorischer und sensorischer Anomalien auf die Entwicklung des Wahrnehmungsvermögens. Man hofft, falls ein kausaler Zusammenhang festgesetzt werden könnte und man ein Mittel fände, die okulomotorischen Störungen frühzeitig zu behandeln, dass es möglich wäre, der Entwicklung gewisser Störungen der Wahrnehmung vorzubeugen.

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Variations in Perceptual Ability Among Types of Cerebral Palsy*

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In a comparison of groups of cerebral palsied and non-cerebral palsied children, matched for age and intelligence, it was found that a larger number of cerebral palsied children showed low perceptual ability, which, in a few cases, was below that of any of the non-cerebral palsied children. Within the cerebral palsied group, perceptual impairment was found to be associated with spasticity rather than athetosis, and with bilateral and left-sided spasticity rather than right-sided. This indicated that perceptual impairment might be related to right-sided brain injury. Motor and visual handicap appeared to have a contributory rather than a determining effect.

THOSE who teach children with cerebral palsy often find that some of them show varying degrees of perceptual impairment. This may take the form of an inability to put together the parts of a 'manikin', to copy patterns, to grade sticks of different lengths, or even just to draw. These disabilities are remarkable since they are greater than can be accounted for by the degree of the children's physical handicap, although, of course, they may occur also among children whose handicap is too great to allow them to do these tasks. Such disabilities are well known among adults with parietal lesions of the cortex (Critchley 1953), and a number of workers, following the earlier studies of Strauss, have found them to be commoner among brain-damaged children than among those with a familial mental defect.

The investigation of the perceptual abilities of random samples of cerebral palsy children is made very difficult by the sensitivity of most of the usual tests to physical handicap, as a result of which comparatively few studies have been carried out. Most studies have included children with only limited handicaps who had manipulative control of at least one hand. Studies comparing the perceptual abilities of cerebral palsy and non-handicapped children matched for age and intelligence, have generally found the cerebral palsy group to be inferior. However, the amount of overlap found between the ranges of performance levels of the cerebral palsy and control groups indicates either that the difference between their perceptual abilities is in fact small, or that perceptual impairment is limited to a sub-group of cerebral palsy children. Since perceptual impairment has been found among cerebral palsy children, the second alternative must apply. The study of Cruickshank, Bice and Wallen (1957) indicates some differences in the perceptual abilities of athetoids and spastics. They gave figure-ground discrimination tests to 211 spastics, 114 athetoids and 110 controls, and found that, while both cerebral palsy groups had poorer performance levels than the controls, the spastics tended to be poorer than the athetoids. Cruickshank and his co-workers unfortunately did not give exact details of the relative levels of motor handicap and intelligence of these

* This research was supported in part by a grant from the National Spastics Society.

groups. Wood (1955) compared the performance of left and right hemiplegics on various tests of figure-ground discrimination, but did not find any significant differences.

The study reported here was an attempt, by means of further subdivision of cerebral palsy types, to find the type with which perceptual impairment was mostly associated. In a previous paper (Wedell 1960) the differences in perceptual abilities found between the subgroups of cerebral palsy children used were analysed. These are summarised in Table I. In the present paper it is intended to state these differences in outline, and to investigate in detail the relationship of some aspects of cerebral palsy, such as degree of motor handicap, aetiology, etc., with these differences, and also to discuss their neurological implications.

Design of Experiment

Subjects. It was decided to compare subgroups of cerebral palsy children, differentiated on the basis of their observed motor disorder rather than on the aetiology of their condition, since data on the latter were not so generally available. Four subgroups of children were collected—one of athetoids, one of spastics with bilateral motor handicap, and one with left and one with right unilateral motor handicap. This subdivision was chosen because it seemed a natural antecedent for most generally accepted classifications. Cases of cerebral palsy can of course rarely be unreservedly classified even under one or other of these heads. Children frequently show motor disorders of mixed types, and the possibility of minimal motor impairment of the opposite side can sometimes not be excluded in hemiplegics. However, children were selected for each group on the basis of their predominant motor disorder.

Various studies have indicated that the distribution of intelligence varies among different subgroups of cerebral palsy children (Woods 1957, Schonell 1956, Perlstein and Hood 1955). To make all the groups

TABLE I—SUMMARY OF GROUPS' SCORES ON TESTS

		Control	*Athetoid*	*Combined Spastic*	*Bilateral*	*Spastic Left*	*Right*
No. of Cases ..		40	20	53	20	13	20
Matching Test	Mean	16.45(*1*)	16.85	15.79	16.25	13.77(*1*)(*2*)	16 65(*2*)
	S.D.	2.93	3.14	3.55	2.20	4.15	2.96
Tracing Test	Mean	10.28	11.0	9.23	8.70	8.38	10.30
	S.D.	4.56	4.99	4.86	4.86	4.98	4.58
Bricks Test	Mean	3.55	3.42	4.55	5.75	5.92(*3*)	2.45(*3*)
	S.D.	3.60	3.63	5.19	5.52	6.43	2 64
Face Test	Freq.	24	15	26	6	7	13
	Prop.	0.60	0.75	0.49	0.30	0.54	0.65
Man. A	Freq.	39	19	40	16	7	17
	Prop.	0.98(*4*)(*7*)	0.95	0.77(*7*)	0.80	0.54(*4*)	0.85
Man. B	Freq.	19	11	11	1	1	9
	Prop.	0.48(*5*)(*6*)(*9*)	0.55(*8*)	0.21(*8*)(*9*)	0.05(*5*)	0.08(*6*)	0.45

(*1*) Difference significant P 0.05 ('*t*' and Wilcoxon's '*T*')
(*2*) ,, ,, P 0.05 ('*t*')
(*3*) ,, ,, P 0.05 ('*t*' and Wilcoxon's '*T*')
(*4*) ,, ,, P 0.001 S.E. = 0.108
(*5*) ,, ,, P 0.01 S.E. = 0.129
(*6*) ,, ,, P 0.05 S.E. = 0.155
(*7*) ,, ,, P 0.01 S.E. = 0.075
(*8*) ,, ,, P 0.05 S.E. = 0.120
(*9*) ,, ,, P 0.05 S.E. = 0.098

comparable in respect of age and intelligence, subjects for each group were selected to fit a predetermined intelligence-age matrix consisting of four 15-point I.Q. brackets between I.Q. 60–120 at five successive ages from 6 to 10 years. The design thus required 20 subjects for each group—one subject for each cell of the matrix. A control group of 40 non-brain-injured children without motor handicap was selected in the same way to provide two subjects for each cell of the matrix. The I.Q. assessments were based on the Stanford-Binet Test. Here again, the possibility of inaccurate assessment of the cerebral palsy children has to be borne in mind, but errors due to a generally known objective procedure seemed preferable to those involved in more subjective forms of assessment. It was in any case not supposed that this method of selection of subjects would allow comparison between individuals in each group.

Subjects were selected from the records of clinics and schools for spastics in various parts of the country. Although the total population from which selection was made approached 2,000 children, it was not possible at the time to find subjects for seven cells of the left unilateral spastic group, and 5 subjects whose I.Q.'s fell slightly outside the appropriate brackets had to be included in other groups. There were, however, no significant differences between the mean chronological ages, mental ages and I.Q.'s of all the groups.

It might be asked whether samples selected in this way were themselves representative of the cerebral palsy population as a whole. It is clear that these samples are not representative in either age or I.Q. range, but since there was no other limitation on the selection of subjects, and since the first appropriate available subject was always used, the samples can be claimed to be random in every other way.

Additional Data. The following data were collected:

Classification of general physical handicap. Schonell's classification, described in Schonell (1956) was used. This consists of 4 categories, ranging from slight (A) to very severe handicap (D). In the present study there were no subjects in handicap category D.

Visual defect. This was subdivided into (*a*) visual defect (specific defect of retina or lens), (*b*) muscular imbalance (e.g., squint) and (*c*) probable monocular vision.

Aetiology. Data were collected under the following 8 categories: (1) Familial. (2) Premature. (3) Neonatal asphyxia. (4) Neonatal jaundice. (5) Post-convulsive. (6) Post-infective. (7) Birth trauma. (8) Unknown.

Birth-weight.

Hand and eye dominance. Hand dominance was assessed by observation, and eye dominance by choice of eye used to look at a picture through a hole in a screen (most frequent of three trials).

I am grateful to Dr. Grace Woods for advice in planning the selection of the data. Apart from hand and eye dominance, these were kindly supplied by the paediatricians concerned with the children.

Tests. Full details of the tests used are given in Wedell (1960). The aim of the selection of the test battery was to use tests of as wide a variety of perceptual tasks as possible, with minimum sensitivity to motor handicaps. The tests used were in part specially constructed and in part adaptations of other tests, and consisted of the following:

1. *Matching Test.* A multiple-choice pattern-matching test, intended to test two-dimensional spatial discrimination.

2. *Light Tracing Test.* A pattern-recognition test including some of the same patterns, in which the pattern to be recognised was 'drawn' on a milk glass screen with a moving dot of light. Level of performance on this test had in a previous study been found to be correlated with ability to draw the patterns.

3. *Bricks Test.* Here the subject had to copy simple brick models. This test was intended to assess three-dimensional spatial discrimination.

4. *Manikin Test.* The Collins Drever Manikin was used. This consists of six pieces of wood cut out in the shape of a head, torso, arms and legs which the subject has to put together appropriately.

5. *Face Construction Test.* The subject was required to put eyes, eyebrows, nose and mouth in their appropriate positions on a blank face.*

This and the Manikin test were included to represent a free-constructional task, which did not involve copying.

Tests 1 and 2 were scored on the number of correct matchings, and test 3 on the number of bricks incorrectly placed—i.e., on the number of errors made. Test 4 was scored on two pass-or-fail criteria, one (B) requiring a perfect performance, and one (A) requiring only a 'human-like' performance. Test 5 was scored on one pass-or-fail criterion of perfect performance. Details of the scoring criteria used will be found in Wedell (1960). Inaccuracies due to lack of motor control were allowed for in the scoring. The only help given to the subjects with more severe handicaps was to put the pieces used in tests 3 to 5 into their hands and to hold down already placed pieces.

Results

Comparisons of the performance levels of the groups were made between each of the cerebral palsy subgroups and the control group, between the combined spastic group and the control and athetoid groups, and between the left and right spastic groups. From Table I it can be seen that the control and athetoid groups were consistently better than the combined spastic group. Also, when the mean scores of the individual spastic subgroups were considered, the performance levels of the bilateral and left spastic groups were found to be consistently worse than those of any of the other groups.

The significance of the differences between the mean scores was assessed both by parametric (Student's '*t*') and by non-parametric (Wilcoxon's '*T*') methods, since the distributions of scores were in some cases skewed.

The differences between the control and combined spastic groups were significant in two instances, between the control and bilateral spastic in one, and between the control and left spastic group in three. The athetoid group was significantly better than the combined spastic group on one test, and the right spastic significantly better than the left spastic group also on one test.

The types of spasticity found among the subjects in the spastic groups are set out in Table II. The distribution of degree of motor handicap and types of visual handicap are shown in Table III for each of the cerebral palsy groups. The bilateral and athetoid groups both contain more subjects in the more severe motor handicap categories, and more subjects with ocular defects of different kinds. The low scoring group referred to in this table contains those subjects whose performance on the perceptual tests was consistently low. This group was selected on criteria of poor performance which varied with mental age

* Derived from the late Mme. Stella Albitreccia, of the *Aide aux Enfants Paralysés, Paris.*

TABLE II—DISTRIBUTION OF TYPES OF SPASTICITY IN SPASTIC SUBGROUPS

Types	*Subgroups*		
	Bilateral Spastic	*Right Spastic*	*Left Spastic*
Quadriplegias ..	13	—	—
Paraplegias	5	—	—
Diplegias	2	—	—
Hemiplegias	—	15	13
Monoplegias ..	—	5	—
TOTALS ..	20	20	13

TABLE III—DISTRIBUTION OF MOTOR AND VISUAL HANDICAP AMONG THE CEREBRAL PALSY GROUPS AND THE LOW-SCORING GROUP

	Athetoid	*Bilateral*	*Spastic Left*	*Right*	*Low-Scoring Group (Prop.)*	*Combined C.P. Group (Prop.)*
No. of Cases	20	20	13	20	14	73
Motor Handicap:						
A Slight	10	9	13	20	0.64	0.71
B Moderate	5	7	0	0	0.13	0.16
C Severe	5	4	0	0	0.21	0.12
Visual Handicap:						
Visual defect	0	4	4	3	0.21	0.15
Muscular imbalance ..	13	12	4	3	0.71	0.44
Monocular vision ..	7	9	3	2	0.50	0.29

in the case of the Matching, Light Tracing and Bricks Tests, but not in the case of the Manikin and Face Tests. If the distribution of motor and visual handicaps among the members of this group is compared with that for the cerebral palsy group as a whole, it can be seen that a larger proportion of subjects in the low-scoring group fall into the severe motor handicap category and into all the visual handicap categories.

Table IV shows the number of subjects in each aetiological category whose level of performance was above the median score on two sample tests, the Light Tracing and Bricks Tests. Subjects whose condition was due to a combination of aetiologies are listed separately at the foot of the table, and grouped under the aetiologies they have in common. These common aetiologies were prematurity, neonatal asphyxia and neonatal jaundice, and in each case the subjects whose condition was due solely to one of these were also included in the numbers given.

The number of subjects whose condition was due to one particular cause was too small to allow for adequate comparison of perceptual performance. However, the proportions of subjects with any one type of aetiology whose performance levels were above the median scores on the two tests were largely similar.

In the figure the proportions of subjects whose levels of performance were above the median scores for the combined cere-

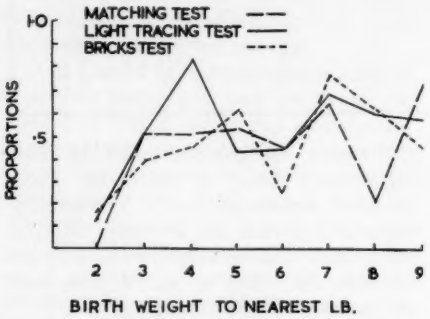
TABLE IV—DISTRIBUTION OF AETIOLOGICAL CATEGORIES AMONG CEREBRAL PALSY SUBJECTS

Frequencies and Proportions of Subjects Scoring Over Median Scores of Light Tracing and Bricks Tests

Aetiological Category	*No. of Cases*	*Light Tracing Test*		*Bricks Test*	
		No. over Median	*Prop.*	*No. over Median*	*Prop.*
1. Familial	4	1	0.25	0	0
2. Premature	17	9	0.53	7	0.41
3. Neonatal asphyxia ..	12	7	0.58	7	0.58
4. Neonatal jaundice ..	3	2	0.67	2	0.67
5. Postconvulsive ..	6	4	0.67	3	0.50
6. Postinfective ..	5	3	0.60	3	0.60
7. Birth trauma ..	7	4	0.56	4	0.56
8. Unknown	7	3	0.43	4	0.56
2 + (2, 4) (2, 3) (2, 3, 4)	27	14	0.52	9	0.35*
3 + (2, 3) (7, 3) ..	18	10	0.56	9	0.50
4 + (2, 4) (2, 3, 4) (4, 3)	9	6	0.67	4	0.50†

* N = 26
† N = 8

bral palsy group on the Matching, Light Tracing and Bricks Tests are shown for successive birth-weights. The feature of this graph which is worth noting is the increase with birth-weight of the proportions at the lowest three birth-weights. At higher birth-weights, the trend becomes irregular.



Proportions of subjects with performance levels on three tests above the median scores for the combined cerebral palsy group, shown for successive birth-weights.

Discussion

The comparisons of the performance levels of the groups show relatively few statistically significant differences. On the other hand, weight is lent to the differences which were found by the fact that the performance level of the combined spastic group was *consistently* below that of the control and athetoid groups, and the bilateral and left spastic groups' performance levels were consistently below those of all the other groups. The question therefore arises whether the design of the study tended to reduce the differences between the groups. This question is dealt with more fully in another paper (Wedell 1960), but the relevant points will be summarised here. Firstly, the wide ranges of age and I.Q. of the subjects in each group produce wide ranges of performance levels, which reduce the statistical significance of the differences found between the means. Secondly, the differences between the groups were found to vary for subjects at successive mental and chronological ages. This was partly due to different developmental rates, and partly to the lower discrimination power of the Matching and Bricks Tests at the higher mental and chronological ages. Lastly, since the Stanford-Binet Test, by which the groups were matched for intelligence, itself contains some perceptual items, the subjects were, to a certain degree, matched for perceptual ability from the start.

In the presence of so many contrary tendencies, the differences found between the groups therefore gain in significance. This necessitates an explanation of the almost invariable, yet statistically insignificant, superiority of the athetoid group over the control group. This appears to be due to the combined effect of two causes. First, the mean I.Q. of the athetoid group is slightly higher than the control group's; this difference again does not approach significance. Secondly, there is a tendency for the intelligence assessments of athetoids to be underestimated on test performance (Saunders 1959), and perceptual ability is, of course, related to intelligence.

So far as the relative performance levels of the athetoid and combined spastic group are concerned, these results thus support Cruickshank's. The finding of a difference between the left and right spastic groups does not agree with Wood's (1955) results.

Table III shows that the relative performance levels of the cerebral palsy groups are not related to the degree of general motor or visual handicap of the subjects in them. Correlations between motor and visual handicap and performance on the tests which were likely to be affected by them were calculated for the combined cerebral palsy group, using Kendall's tau_c. Although the coefficients were positive for almost all tests, none were significant. Performance on the Bricks Test came nearest to being affected by general motor handicap (tau_c 0.207). This is interesting, since this was the only test on which the athetoid group's performance was poorer than the control group's. Of the three forms of visual handicap, muscular imbalance

most affected Matching Test performance (tau_c 0.128) and monocular vision most affected Light Tracing Test performance (tau_c 0.20). It seems, then, that both motor and visual handicap contribute to perceptual impairment but are not sufficient to determine it.

Floyer (1955) in her study of cerebral palsy children in Liverpool, came to a similar conclusion. Wood (1955) related several measures of visual defect to performance on figure-background discrimination tests, but found only that subjects with poor visual acuity tended to perseverate.

The relationship between the various forms of aetiology and performance on perceptual tasks could unfortunately not be adequately established on the data available. Apart from the 'familial' subjects, those with an aetiology of prematurity have slightly smaller proportions of subjects making above-median scores. The relationship between prematurity and level of performance comes out more clearly in the figure. There is an indication here that degree of prematurity is related to perceptual performance. Performance on the Light Tracing and Bricks Tests was correlated with birth-weight for subjects with birth-weights of 5 lb. and below, but the coefficients, although positive, did not reach significance (tau_c 0.238 and 0.230, respectively). When the mean I.Q.s of subjects at successive birth-weights were plotted, the graph closely followed those shown in the figure, but fell further at the higher birth-weights. This supports similar findings reported by Fraser-Roberts and Asher (1949). Confirmation of the relationships indicated clearly requires a larger sample. The differences in test performance found between the cerebral palsy subgroups were not found to be related to the proportions of subjects in each group with low birth-weights.

As a form of cross-validation for the differences in performance levels found between the groups, the low-scoring group of subjects was set up as described above. This group was then found to contain 7 subjects of the bilateral, 4 of the left and 1 of the right spastic groups, and 2 of the athetoid group as well as 3 subjects of the control group. These results largely parallel the differences found between the complete groups. The higher incidence of motor and visual handicap in this group brings out again the contribution of both these factors to perceptual impairment.

The proportions of subjects with birth-weights of 5 lb. and below were 0.54 in the low-scoring group and 0.42 in the combined cerebral palsy group. All types of aetiology were represented among the subjects of the low-scoring group. Those selection criteria which did not vary with mental age unavoidably tended to sort out the younger rather than the older subjects, but levels of intelligence were evenly distributed in the low-scoring group. The I.Q.s of the three subjects of the control group, however, were all below 75.

Prematurity, degree of physical handicap, and ocular defect thus all appear to contribute to perceptual impairment but cannot be regarded, on the present data, as direct causes of it. Prematurity no doubt exerts an influence through the damage to the central nervous system with which it is often directly or indirectly associated. General motor handicap and ocular defect may impede the child's acquisition of sensory motor experience, which forms an essential groundwork for visuo-spatial abilities. In addition, the possibility cannot be ruled out that the more severe forms of these handicaps did affect performance on the tests in this study, although their selective effect on the bilateral spastic as opposed to the athetoid subjects would then require explanation.

The results indicate more clearly, however, that type of cerebral palsy is the main determining factor in perceptual impairment. The superiority of the athetoid to the combined spastic group indicates, not surprisingly, that the cortical injury involved in spasticity is a likely cause. The low performance of the bilateral spastics is thus easily explained, but the difference in performance between the right and left

spastic groups goes against present views on the plasticity of cortical function in young children. As has already been mentioned, Wood (1955) found no difference in figure-ground discrimination between left and right hemiplegics, using groups with a wider age-range. On the other hand, in a subsequent study (Wood 1959) she found a non-significantly lower mean I.Q. among her left hemiplegics. Perlstein and Hood (1955) found no significant difference between the I.Q.s of their left and right hemiplegics, while Woods (1957) found a larger proportion of ineducable children among the left hemiplegics.

The finding of lower perceptual ability among the left spastics in the present study parallels the findings among adults with right (minor hemisphere) parietal lesions (e.g., McFie *et al.* 1950). The dominance of the supposedly affected hemisphere in the left spastic group is hard to establish. It is rarely possible to distinguish between congenital and acquired hand or eye dominance in cerebral palsy children. If the available data are used, the association of perceptual impairment with lesions of the minor hemisphere is indicated on the basis of hand dominance. Eye dominance gives conflicting results. Ipsilateral hand and eye dominance tends to support the indications of hand dominance.

The data for the left spastic group therefore conform closely to those found among adults. This fact, and Wood's (1957) findings, provide a context for the present results. None the less, the contrary evidence mentioned above allows them only to be put forward tentatively at present, while further work is carried out to verify them.

SUMMARY

Four groups of cerebral palsy children (one of athetoids, and one each of spastics with bilateral, left-sided and right-sided motor defect) were compared with each other and with a control group of non-brain-injured children without motor handicap on several perceptual tests. All five groups were matched for age and intelligence.

The age and I.Q. ranges of all groups were from 6 to 10 years and from I.Q. 60 to 120.

The control, athetoid and right spastic groups' levels of performance were not significantly different, but those of the bilateral and left-sided spastic groups were consistently lower.

Motor and visual handicaps were found to have a slight effect on perceptual ability.

A possible relationship between perceptual impairment and degree of prematurity was indicated.

The neurological implications of these findings are discussed.

RÉSUMÉ

Variations dans la faculté de perception propre á certains types d'infirmité motrice cérébrale.

Quatre groupes d'enfants infirmes moteurs cérébraux (athétoïdes, spastiques avec déficience motrice bilatérale, gauche et droite) ont été comparés entre eux et par rapport à un groupe témoin d'enfants sans lésion cérébrale et sans infirmité motrice. Les cinq groupes étaient équivalents en âge et en intelligence.

L'âge de tous les groupes variait entre 6 et 10 ans, le Q.I. entre 60 et 120.

Les degrés de performance relevés dans le groupe témoin, le groupe des athétoïdes et celui des spastiques droits n'ont pas été sensiblement différents alors que ceux relevés dans le groupe des spastiques bilatéraux et gauches ont été uniformément plus bas.

On a trouvé que les infirmités motrices et visuelles avaient une faible répercussion sur la faculté de perception.

On a signalé la possibilité d'un rapport entre l'inaptitude à percevoir et le degré de prématurité.

Les conséquences neurologiques de ces constatations sont discutées.

ZUSAMMENFASSUNG

Unterschiede im Wahrnehmungsvermögen zwischen einigen Typen der zerebralen Kinderlähmung.

Vier Gruppen von Kindern mit zerebraler Lähmung (eine von Athetoiden und je eine von Spastikern mit bilateraler, linker und rechter motorischer Störung) sind untereinander und mit einer Kontrollgruppe von Kindern mit ungeschädigtem Gehirn und ohne motorische Behinderung verglichen worden. Alle fünf Gruppen waren aus Kindern gleichen Alters und gleicher Intelligenz zusammengesetzt.

In allen Gruppen schwankte das Alter zwischen 6 und 10 Jahren und der I.Q. zwischen 60 und 120.

Die Leistungshöhen der Kontrollgruppe, und der Gruppen der Athetoiden und rechten Spastiker waren nicht bedeutsam verschieden, während die der Gruppen der bilateralen und linken Spastiker einförmig niedriger waren.

Man hat gefunden, dass motorische Behinderungen und Sehstörungen eine mässige Wirkung auf das Wahrnehmungsvermögen haben.

Es ist auf die Möglichkeit einer Beziehung zwischen der Beeinträchtigung des Wahrnehmens und dem Grade von Frühgeburt gewiesen worden.

Die neurologischen Folgerungen dieser Befunde sind besprochen worden.

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The Hypotonic Forms of Cerebral Palsy

AN ANALYSIS OF 41 PERSONAL CASES AND AN ATTEMPT TO EXPLAIN THEIR ORIGIN

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Introduction

FREUD had previously drawn attention to the ataxic components in some children with encephalopathies of perinatal origin, but it was Foerster in 1909 who first described a form of cerebral palsy in which hypotonia and ataxia were the special features. Foerster termed this disorder the 'astatic type of cerebral infantile paralysis'. The patients had an abnormal perinatal history. Besides their greatly diminished muscle tone, they also often showed strabismus, extensor plantar responses, epileptic fits and oligophrenia, clinical features which are common in the classical forms of cerebral palsy described by Little and whose presence here illustrated the close relationship between the hypotonic and other forms of cerebral palsy.

Foerster believed that the hypotonia and ataxia resulted from birth damage to the cerebellum, and more specifically from intracerebellar bleeding during labour. Postmortem evidence of damage to the cerebellum in one such patient was described later by Batten and von Wyss (1915), but in his own two cases Foerster found no changes in the external appearance of the cerebellum but extensive atrophy in the frontal regions of the cerebrum. De Capite (1925) attributed hypotonia of central origin in young children to disorders in the lenticular nucleus. The case described by Marinesco and Draganesco (1929) also had changes in the basal ganglia as well as in the cortex and pyramidal tracts, but the presence of chorea and other clinical abnormalities suggested that there might be extrapyramidal involvement in this case.

Clinically the hypotonic form of cerebral palsy is characterised by there being diminished muscle tone. The patients show an increased range of passive movement in their joints, particularly in the legs. It is easy to put their feet behind their head, or their big toes in contact with the nose. They can be rolled into a ball with their head between their feet, which we propose to call the 'armadillo sign' (Fig. 1), and their arms can be wound round their throat

Fig. 1. Hypotonic cerebral palsy. Bending the trunk. The 'armadillo sign'.

like a scarf, which we call the 'scarf sign'—the 'signe de foulard' of the French authors.

The power of voluntary movement and resistance to passive movement are diminished, but the tendon jerks tend to be brisker than in cases of atonia of spinal origin. The patients find great difficulty in standing or walking and very easily fall over. Their gait is often ataxic, the ataxia being of the cerebellar type in some cases and more like the ataxia of spinal cord disease in others.

Ford (1952) describes an ingenious way of distinguishing the hypotonic form of cerebral palsy from spinal amyotonia (Oppenheim's disease) in very young children. When the amyotonic child is held erect the tone of his legs remains poor and they hang loosely. On the other hand, a child with hypotonia arising in the brain flexes his legs like a healthy infant (Fig. 2), even though their tone when he lies down may be as poor as in the amyotonic child. (This is sometimes called Foerster's sign.)

Fig. 2. When the child with hypotonic cerebral palsy is held up, the legs flex like a healthy baby's, in contrast with spinal amyotonia, where they hang down loosely.

In 1952, Yannet and Horton described 31 children with hypotonic cerebral palsy. They divided them into atonic, athetoid and ataxic types, all the children being severely retarded mentally. The 15 atonic cases had greatly increased tendon reflexes and a third of them showed extensor plantar responses. (The cases described include a Tay-Sachs and a tuberose sclerosis.) The 10 ataxic cases included two with probable kernicterus from neonatal jaundice. The I.Q. of the children in the ataxic hypotonic cerebral palsy group averaged 35, compared with 15 for the atonic group and 20 for the six athetoid cases. The severity of the intellectual defect, the high incidence of fits, and the tendency to microcephaly point to widespread pathological changes in the hypotonic cerebral palsies. According to Yannet and Horton, hypotonic cerebral palsies may be caused by agents acting before, during or after birth.

Other writers on cerebral palsy have tended to neglect the hypotonic form. It is not rare, however, and we determined to make a detailed study of it.

Personal Observations

Case Material and Methods of Examination

Our series of 41 patients comprises 21 children from the Neurological Clinic and 21 from the Institution for the Re-education of Cerebral Palsied Children in Zeleznice, one patient being common to both groups. The diagnosis of hypotonic cerebral palsy was established in all cases, patients in whom it was in doubt being excluded from consideration. There were 16 girls and 25 boys, their ages ranging from 11 months to 11 years. They form the largest series of cases of atonic cerebral palsy so far published.

The cases were studied from various aspects. The histories were analysed from the point of view of elucidating aetiological factors, assessing the rate of the child's

motor and mental development, and determining the presence or absence of epilepsy. In our neurological examinations we searched for evidence of pyramidal, extrapyramidal and cerebellar disease, and tried to assess the child's mental state. Other investigations included electroencephalography, examination of the cerebrospinal fluid, and especially pneumoencephalography.

Findings

The causative factors in the 33 patients in whom these could be elucidated differed little from those we reported in 1956 in a series of 204 patients suffering from a variety of different types of cerebral palsy (Lesny, Dittrich *et al.* 1956) though slightly more of the present series were prematurely born.

Rather more than half the patients (22) were oligophrenic,* and delayed development was noted in 36 (88 per cent). One or more epileptic fits had occurred in 10 (25 per cent), but only 6 (14 per cent) had frequent seizures.

The most interesting finding was the relative rarity of signs of cerebellar dysfunction, even when these were looked for with care. Since only 13 (32 per cent or less than a third) showed any indication of cerebellar abnormality, hypotonic cerebral palsy should not be assumed to be necessarily or always cerebellar in origin, though cerebellar signs are sometimes present.

Extrapyramidal hyperkinesias were found in 11 patients (27 per cent) and these were usually choreic in type. Unwanted movements are thus relatively frequent in hypotonic cerebral palsy.

The tendon-jerks were normal in 23 cases (57 per cent), but in the remainder almost all possible combinations of abnormal jerks were noted. They were exaggerated in both arms and legs in 8 cases (20 per cent). Signs indicating pyramidal tract damage occurred so commonly that they were considered part of the characteristic clinical picture. They were found in 24 cases (57 per cent). There was only one case of spastic hemiplegia and one of paraplegia, these apparently representing clinical pictures of cerebral palsy that are transitional between the atonic form and other types. We did not estimate the chronaxie in these cases, for this question has been dealt with by Lesny, Odvárková and Stein (1956). They showed that in contrast to other types of cerebral palsy, the chronaxie is lengthened in hypotonic cerebral palsy, the cathodic chronaxie being more affected than the anodic. In atonic cerebral palsy the chronaxie was usually between 1.5 and 3 millisec. This may be compared to the normal upper limit of 0.9 millisec in unaffected children over twelve months of age.

The electromyographic findings are more abnormal in hypotonic cerebral palsy than in any other type. The spikes are narrowed, showing a reduction in duration from the normal of about 10 to 1.3 milliseconds and a lowering in voltage, findings similar to those obtained in some diseases of muscle.

An EEG was done in 40 cases. The findings are shown in the table and illustrated in Figs 3 and 4. In 5 cases the EEG was normal; in 11 there was a generalised abnormality; 6 cases showed focal parietal or biparietal abnormality; 6 showed synchronous episodic activity with sharp waves, 3 of these cases also having clinical fits. In 5 cases there were spikes and waves, 4 of these having clinical fits. In the remaining 7 cases dominant fast activity was observed; in some of these the EEG could be considered 'hypermature'. In 2 of these last 7 cases the child had clinical fits.

Pneumoencephalography was performed in 18 patients. In one of these the result was normal. In the remaining 17 patients various abnormalities were encountered, including differences in the sizes of the ventricles and defective filling of the subarachnoid space and basal cisterns. In 2 cases there was cerebral atrophy, unilateral in one and bilateral in the other.

Progress under Observation

Rehabilitation without drug treatment was applied in 18 cases and only 3 of these

* I.e. with an I.Q. under 60 per cent.

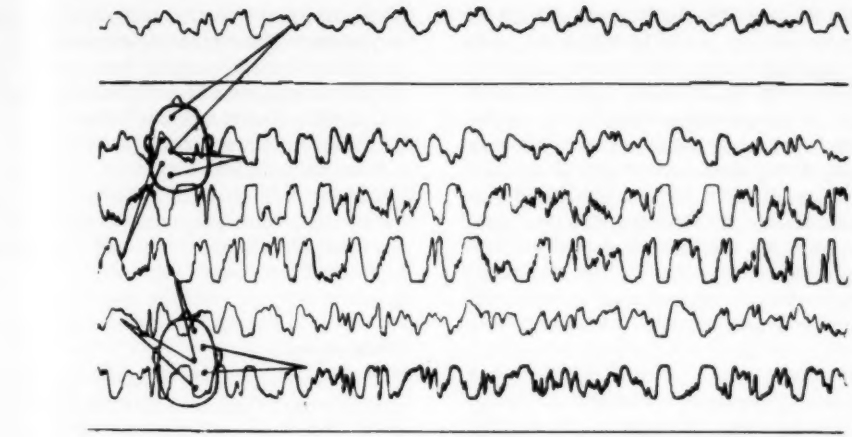

Fig. 3. (*above*) EEG of girl of 5 years. Hypotonic cerebral palsy with epileptic fits. Generalised dysrhythmia; slow waves with some superimposed fast activity.

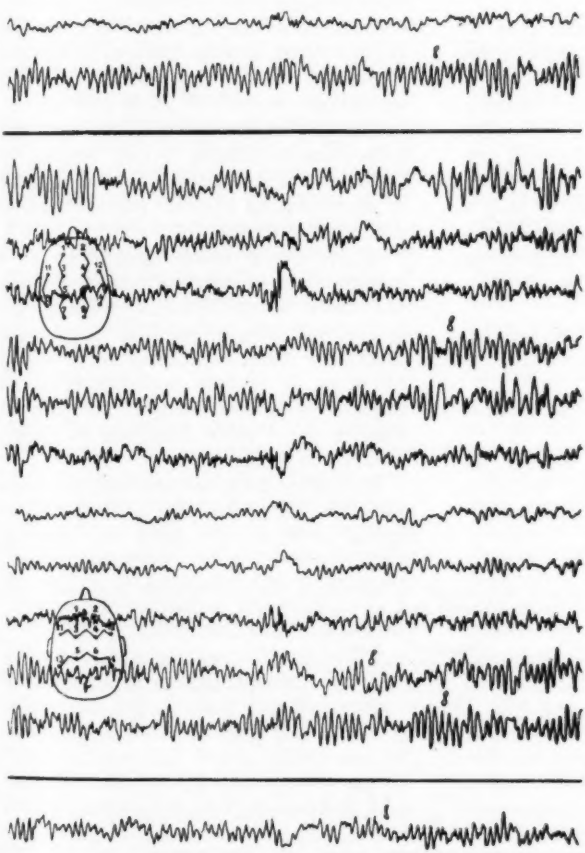

Fig. 4. EEG of boy of 17 months. Hypotonic cerebral palsy. Dominant activity 8/sec. Hypermature EEG.

(16.5 per cent) did not improve—about the proportion that would be expected in spastic cerebral palsy. There were a similar number of spontaneous improvements, again corresponding roughly to the proportion expected in other forms of cerebral palsy. Drug treatment improved all the 3 patients given it, but it is impossible to evaluate this observation. Pituitary gland implantation was done in 6 patients, 5 of whom improved after it. An account of this procedure will be found in an accompanying article (Lesny, Vojta and Klen, 1960).

Present Observations in Brief

We can summarise our observations as follows:

(1) Aetiologically, the hypotonic form of cerebral palsy does not differ from other forms.
(2) The same is true of associated epilepsy.
(3) Signs of pyramidal tract disorders are found in rather more than half the cases.
(4) Cerebellar signs are found in only a third of the patients. Patients with hypotonic cerebral palsy showing cerebellar signs did not differ in other respects from those without them.
(5) Extrapyramidal hyperkinesias are present in just over a quarter of the patients.
(6) The cerebrospinal fluid is usually normal.
(7) Pneumoencephalography almost always reveals abnormalities, but there are no typical pneumoencephalographic findings.
(8) Intellectual defect is frequent among patients with hypotonic cerebral palsy. Delayed development was noted in 88 per cent, and 54 per cent were oligophrenic. This is a higher frequency than is seen in other forms of cerebral palsy, except tetraplegia.
(9) There are gross EEG abnormalities of the generalised type, sometimes of epileptic nature, even when fits are absent.

Discussion

The hypotonic form of cerebral palsy is certainly not a disease entity to be separated from other forms of cerebral palsy, for it has too many clinical findings in common with them. Its only characteristic features are hypotonia, and the frequent occurrence of serious mental impairment, either of which may be part of other clinical syndromes in cerebral palsy. The hypotonic form attracts interest largely because of difficulties in interpreting the significance of the hypotonia in either physiological or pathological terms.

Hypotonia is much commoner as a manifestation of nervous disease in children than in adults. It is not only a feature of cerebral palsy of prenatal or perinatal origin, for it has been observed in infants of 10 or 12 months suffering from the sequelae of various kinds of encephalitis. One of the 4 cases described by Orrico (1927) was of this type.

Hypotonia does not necessarily arise in the brain. Hypotonia due to disease of the spinal cord in the form of Werdnig-

ELECTROENCEPHALOGRAPHIC FINDINGS IN 40 CASES OF CEREBRAL PALSY, SHOWING THE CHILDREN'S INITIALS AND AGE IN YEARS

Type of Cerebral Palsy	*Generalised Abnormality With Synchronization*	*Generalised Abnormality Without Synchronization*	*Focal Signs*	*Generalised Spikes and Waves*	*Spikes or Sharp Waves*	*Normal*	*Fast Activity*	*Hypermature EEG*	*Biparietal Abnormality*
Hypotonic	P, 2½	M, 12 G, 7 J, 4 K, 6 C, 4 K, 6 B, 5 H, 6 T, 7 M, 7½	D, 4 B, 3½ K, 4	S, 6	H, 4 J, 1½ K, 6	R, 6 H, 3½ T, 2 T, 2 K, 7	T, 2½ S, 7	V, 6 N, 1½ N, 7	T, 4 S, 3½ C, 12½
Hypotonic with fits	—	—	—	T, 7 B, 6 P, 6½ H, 6	T, 9 P, 3½ C, 2	—	P, 1½ Z, 6	—	—

Hoffmann's disease or Oppenheim's amyotonia congenita is more widely known than hypotonia of cerebral origin. In Oppenheim's disease there is a congenital developmental defect of the anterior horn cells of the spinal cord, while in Werdnig-Hoffmann's disease these cells show a progressive degeneration. The damage to the anterior horn cells, which in the adult leads to flaccid paralysis with muscular atrophy, gives rise to hypotonia in the child.

One of our patients showed a change from hypotonia to hypertonia during an intercurrent bronchopneumonia, with a return to the original hypotonia when the infection was over. A similar variation of tone has been observed by Byers (1941). He stated that weakness and hypotonia precede spasticity in the hemiplegic form of cerebral palsy, and he ascribed these symptoms to cortical damage during birth.

Its frequent appearance in early childhood gives hypotonia an importance in the neurology of this stage of life. As a localising sign of nervous disease, in early childhood the value of hypotonia is impaired because it is associated with lesions in various sites. Furthermore, this is a sign that we have, so to speak, to evaluate in time as well as in space. *Hypotonia in the child is a finding to be assessed in terms of maturation as well as of localisation in the nervous system.*

In child neurology there are signs indicating the site of the disorder in the nervous system, and there are also developmental signs determined by the level of maturation. A knowledge of the developmental signs and skills of each level of maturation in childhood is essential if *developmental signs* are not to be misinterpreted as *localising signs* in the individual patient.

It is not possible, even with our recently changed ideas of the neurophysiological mechanisms governing muscle tone, to do more than speculate on why hypotonia is relatively common in early childhood. As Pitha, Masin and Polak (1956) remark, disorders of the afferent as well as the efferent pathways must be considered when discussing the possible causes of disorders of muscle tonus. The parietal lobes must also be considered.

Clinical experience supports this opinion, for diminished muscle tone often accompanies disorders of sensation, as in tabes dorsalis, Friedreich's ataxia, and other diseases in which disturbances of sensation due to disorders of the afferent arc are of equal or even greater importance than those affecting the motor pathways. In hypotonic cerebral palsy we see ataxia of the posterior column type at least as often as ataxia of the cerebellar type.

It seems reasonable to speculate whether hypotonia in cerebral palsy might be a result of functional immaturity of the afferent side of the child's nervous system, or might be caused by damage to the sensitive analysers of afferent stimuli in the parietal region.

In early childhood only diffuse sensibility is present. Discriminative perception develops gradually and may be demonstrated by the age of 3 or 4 years. The immaturity of the afferent nervous system before this age is probably the reason why hypotonia is produced more readily then than later. Thus while parietal lobe damage may produce hypotonia in older children and adults, lesions in a wide variety of situations may produce it in younger children.

Further evidence in favour of this concept is provided by the electro-diagnostic findings already mentioned. Chronaxie is lengthened much more significantly in diseases of the afferent neurone than in cerebral motor lesions, in which the reduction in excitability is relatively slight. Thus chronaxie is lengthened in Friedreich's disease, tabes dorsalis, etc. It is also lengthened in hypotonic cerebral palsy, and in 20 cases tested it was found to be much longer (from 2 to 10 millisec) than in 10 control cases of spastic cerebral palsy (0.3–1 millisec).

The electromyographic changes described do not exclude disorders of the afferent system, for it is known that disorders of the afferent arc may affect the electromyogram. Moreover, 6 patients who were studied showed discharges of higher

voltage on maximal contraction than on contraction against resistance. This observation has been described by Isch (1957) as characteristic of disorders of deep sensibility.

There is some pathological evidence to support the view that the hypotonic syndrome may be caused by lesions in the afferent system. In Foerster's two cases the frontal atrophy spread backwards to the central gyri and into the parietal region. In the case reported by Marinesco and Draganesco (1929a) cell changes were found in the third and fourth layer of the cortex of the whole brain. Most of the known histological findings in the hypotonic form of cerebral palsy show such extensive brain involvement that not a great deal can be learnt from them. But Marinesco and Draganesco (1929b) mention a case of this disease which Ramirez-Hernandez demonstrated to them. When in due course he sent the child's brain for pathological examination they found 'cystic degeneration' of the white substance in the left parieto-temporal convolution, gyrus submarginalis and gyrus angularis. Otherwise no gross histological changes in the nervous system were noted. This appears to be the only reported case of the hypotonic form of cerebral palsy in childhood in which neuropathological changes were strictly localised.

Such are our present views of the origin of the hypotonic form of cerebral palsy. We hope they will be corrected, changed and elaborated so that we shall know more of the pathological and physiological processes involved. This may be a step towards improving the outlook for children with this condition, which is the aim of our efforts.

SUMMARY

Forty-one cases of the hypotonic form of cerebral palsy, 16 girls and 25 boys, were carefully examined in hospital practice.

The aetiological factors and prevalence of epileptic seizures were similar to those found in other forms of cerebral palsy, the mental retardation being slightly greater. In the majority, tendon reflexes were normal but plantar responses were extensor. Involuntary movements of extrapyramidal origin, usually choreoid in type, occurred in more than a quarter of the patients. Cerebellar signs were observed in less than a third, which demonstrates that the condition cannot be attributed to cerebellar disease in all cases.

The cerebrospinal fluid was usually normal. Air encephalograms usually showed abnormalities, but these were not characteristic. The electroencephalograms showed generalised abnormalities in 11 of the 40 cases examined.

Our observations show that the hypotonic form is a form of cerebral palsy and not a disease entity. Hypotonia itself is a symptom typical of nervous diesase in early infancy and it may arise from various causes. The site of the lesion may be very different in different cases. Therefore it is necessary to consider hypotonia as a developmental sign of early childhood rather than a specific localising sign.

The probable reason for the frequent occurrence of hypotonia in childhood is that for normal function skeletal muscle requires good afferent innervation, and in infants the afferent system is immature and 'diffuse sensibility' prevails. Against this background, hypotonia may be produced relatively easily at this age by a variety of lesions.

There is some histological evidence for this concept from a few verified cases, especially the Ramirez-Hernandez case, the only example of this type of cerebral palsy in which only one localised lesion was found; here it was situated in the left temporo-parietal region.

RÉSUMÉ

Les formes hypotoniques de l'infirmité motrice cérébrale.

L'étude porte sur 41 cas de forme hypotonique de l'infirmité motrice cérébrale. 16 filles et 25 garçons ont été examinés avec soin à l'hôpital. Les observations sont rapportées:

Similitude avec les autres formes d'infirmité motrice cérébrale, en ce qui concerne les facteurs étiologiques et la prédominance des crises d'épilepsie. Le degré de retard mental un peu plus élevé. Reflexes tendineux normaux, mais signe de Babinski dans la majorité des cas. Mouvements involontaires d'origine extrapyramidale—pratiquement de type choréique—chez plus du quart des malades. Signes cérébelleux chez moins du tiers, preuve évidente que cet état ne peut être attribué dans tous les cas à une maladie du cervelet.

Le liquide céphalorachidien était généralement normal. Les encéphalogrammes gazeux montraient généralement des anomalies mais celles-ci n'étaient pas caractéristiques. Les électroencéphalogrammes montraient des anomalies généralisées dans la moitié des cas examinés.

Nos observations montrent que la forme hypotonique est une forme de l'infirmité motrice cérébrale et non une entité pathologique. L'hypotonie elle-même est un symptôme typique en tant que manifestation de maladie nerveuse de la toute première enfance et ses causes peuvent être diverses. Le siège de la lésion peut ne pas être le même dans différents cas. C'est pourquoi il faut considérer l'hypotonie comme un signe évolutif de la première enfance plutôt qu'un signe de localisation lésionnelle.

Il est probable que la fréquence des cas d'hypotonie dans l'enfance tient au fait que, pour un fonctionnement normal, les muscles du squelette réclament une bonne innervation afférente. Chez les nourrissons, le système afférent est immature et il existe une prépondérance de 'sensibilité diffuse'. Ceci étant, l'hypotonie peut être provoquée assez facilement à cet âge par toute une série de lésions.

Cette conception est corroborée par certaines preuves histologiques trouvées dans quelques cas vérifiés, notamment le cas Ramirez-Hernandez, l'unique exemple de ce type d'infirmité motrice cérébrale où l'on n'a trouvé qu'une seule lésion localisée; dans le cas d'espèce, celle-ci était située dans la région pariéto-temporale gauche.

ZUSAMMENFASSUNG

Hypotonie bei zerebraler Kinderlähmung.

41 Fälle von hypotonischen Formen der Zerebrallähmung, 16 Mädchen und 25 Jungen, sind sorgfältig im Hospital untersucht worden. Die Befunde werden berichtet:

Die ätiologischen Faktoren, das überwiegen der epileptischen Anfälle glichen den Befunden der anderen Formen von Zerebrallähmung, der Grad der Verspätung der geistigen Fähigkeiten war ein bisschen höher. In den meisten Fällen gab es normale Sehnenreflexe aber Fussohlenantwort in Streckung. Unfreiwillige Bewegungen, extrapyramydalen Ursprungs, gewöhnlich choreoider Form, kamen bei mehr als einem Viertel der Kranken vor. Zerebellare Symptome wurden bei weniger als einem Drittel beobachtet, was beweist, dass man diesen Zustand nicht in allen Fällen einer Erkrankung des Kleinhirns zuschreiben kann.

Das Liquor war meistens normal. Die Pneumoenzephalogramme wiesen meistens Abnormitäten auf, die aber nicht charakteristisch waren. Die Elektroenzephalogramme zeigten generalisierte Anomalien bei der Hälfte der untersuchten Fälle.

Unsere Beobachtungen zeigen, dass die hypotonische Form nur eine Form der Zerebrallähmung und nicht eine selbstständige Erkrankung ist. Hypotonie selbst ist ein Symptom, typisch als Kennzeichen einer Nervenkrankheit der ersten Kindheit, und kann durch verschiedene Ursachen hervorgerufen werden. Der Sitz der Schädigung kann sehr verschieden von einem Falle zum Anderen sein. Darum muss man Hypotonie vielmehr wie ein Entwicklungsmerkmal der ersten Kindheit als wie ein spezifisches Lokalisationszeichen betrachten.

Die Häufigkeit der Hypotonie in der Kindheit ist wahrscheinlich dadurch bedingt, dass die Skelettmuskeln, um normal funktionnieren zu können, einer guten afferenten Inner-

vation bedürfen. Beim Säugling ist das afferente System unreif und die 'diffuse Sensibilität' nimmt überhand. Auf diesem Hintergrund kann Hypotonie ziemlich leicht in diesem Alter durch allerlei Ursachen entstehen.

Diese Auffassung wird durch gewisse histologische Beweise bekräftigt, die in einigen kontrollierten Fällen gefunden wurden, besonders in dem Fall Ramirez-Hernandez, einziges Beispiel dieses Typus der Zerebrallähmung, wo nur eine einzige lokalisierte Schädigung aufgefunden wurde, hier im linken Scheitel-Schläfenbezirk.

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Pituitary Implantation in Cerebral Palsied Children

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THERE are few effective medical methods of treating cerebral palsy. Apart from motor re-education there is in fact no useful treatment at all and in many cases re-education is clearly insufficient.

In most cases of cerebral palsy the development of the central nervous system is delayed. The delay mainly affects motor and psychomotor development, but in some cases there is mental retardation also. It seemed that accelerating central nervous system development might help in our efforts to treat cerebral palsy and we therefore determined to use pituitary gland, which has a well-known action on growth and development.

A great deal of work has been done on the implantation of pituitary gland in endocrine, especially pituitary, disorders. Such implantations have been performed in children as well as adults—for example, by Lanezkaja (1951) in diabetes insipidus and Geisler (1954) in various endocrine disorders in children. We decided to try it in children with cerebral palsy.

We used preserved bovine pituitary gland, which was always ordered for a certain day, brought by express train and collected at the railway station. It was transported in a sterile and cooled container. The implantation was performed in the morning of the same day, under the skin of the abdomen, infection being prevented by submerging the implants in saline containing 20,000 units of penicillin and 1 g. of streptomycin per 2 ml.

Since 1955 we have performed pituitary-gland implantation in 58 boys and 26 girls between the ages of 18 months and 5 years; 65 of the 84 were suffering from cerebral palsy and 19 from related conditions.

Cases were considered 'Improved' if in four weeks, without any other treatment, they showed considerable amelioration in mobility, speech, behaviour and general condition. When two or three of these capacities were improved the cases were considered 'Mildly Improved'. When only one of the capacities showed amelioration within four weeks, this was ascribed to chance and the case was recorded as 'Not Improved'.

TABLE I

RESULTS OF IMPLANTATION, JUDGED FOUR WEEKS LATER

Form of Cerebral Palsy	*Improved*	*Mildly Improved*	*Not Improved*
Diplegic	2	8	3
Hemiplegic	1	1	1
Tetraplegic (bilateral hemiplegia)	1	1	3
Hypotonic	10	14	4
Extrapyramidal (athetotic)	2	12	2
Delayed development only	0	0	1
Microcephaly with oligophrenia	0	5	4
Mongoloid oligophrenia (Langdon Down's disease)	3	4	2
TOTALS	19	45	20

The figures in Table I show that pituitary-gland implantation has given good results. Improvement was noted in 22.5 per cent of the cases and mild improvement in 53.5 per cent. The greatest success was obtained in hypotonics, which comprised about a third of our cases. It is the hypotonics who most often need medical treatment before re-education.

Estimation of Hormone Contents

Pituitary implantation clearly improves some cases of cerebral palsy. This improvement was apparent in their greater mobility, a reduction in their increased reflexes, and normalisation of their tonic conditions. Several of the children began to walk after their implantation. In some cases the children did not start to speak and keep themselves clean until after implantation.

It is not easy to explain why the implantation of pituitary gland was successful in some cases, whereas injection of various pituitary hormones has had hardly any effect. In the hope of throwing some light on the problem we tried to ascertain the hormone contents of the pituitary grafts.

The still frozen pituitary glands were unwrapped. The washed glands were then allowed to thaw at room temperature and immediately after thawing they were homogenised in glass homogenators and physiological solution was added to make a total volume of 2 ml. for each pituitary gland. The homogenised material was deprived of ligamentous fibres by filtration through a sterile silk filter. The pituitary tissue homogenised in this way was then injected into animals.

Hormonal activity was determined as follows:

(1) Thyrotrophic hormone (TTH) by the method of Rowland and Parkes (1954) on infant female guineapigs.
(2) Corticotrophic hormone (ACTH) by Sayer's method on pituitectomised rats (Sayers *et al.* 1948).
(3) Somatotrophic hormone (STH) by Evans's method on pituitectomised rats (Li and Evans 1947).
(4) Luteotrophic hormone (LTH) by the method of Riddle on doves (see Lesny *et al.* 1955).

For comparison, we employed partly international standards (ACTH and LTH) and partly our own national standards (TTH and STH).

Our aim was to ascertain the activity of these hormones in bovine pituitary glands, and to follow the effects of freezing on the activity of the hormones during three months' storage. The results are shown in Table II, the figures showing the hormonal activity of one pituitary gland.

TABLE II
EFFECTS OF STORAGE ON HORMONE CONTENTS OF BOVINE PITUITARY GLANDS

	TTH (*Acron units*)	*ACTH* (*int. units*)	*STH* (*Evans's units*)	*LTH* (*micro-units*)
Fresh gland	—	0.6	1.5	10
After 14 days	1	2	1.5	9
After 1 month	1	1	1.5	—
After 3 months	1	—	—	11

It will be seen that bovine pituitary glands show an important activity in TTH, STH and LTH. It is clear that storage up to three months does not reduce the activity of TTH and LTH, and that of STH is not reduced up to one month. The activity of ACTH is very low, so that its significance is negligible.

Conclusion

We feel certain that one of the three hormones mentioned, and perhaps all three of them, can accelerate the delayed development of the central nervous system in cerebral palsied children.

SUMMARY

Implantation of stored bovine pituitary gland was performed in 84 children, 65 of whom were suffering from cerebral palsy and 19 from related conditions. Improvement was observed in 19 cases (22.5 per cent) and slight improvement in 45 cases (53.5 per cent). Titrations of the hormone content of the grafts showed that the thyrotrophic, growth and luteotrophic hormones were present even after a month's storage, whereas the adrenocorticotrophic hormone had disappeared.

RÉSUMÉ

Implantation d'hypophyse chez les enfants infirmes moteurs cérébraux.

84 enfants, dont 65 infirmes moteurs cérébraux et 19 atteints de maladies apparentées, ont été traités par implantation d'hypophyse boving convervée. L'amélioration fut certain dans 19 cas (22,5 pour cent), légère dans 45 autres cas (53,5 pour cent). Les titrages du taux hormonal ont révéle que, même après une durée de conservation d'un mois, les hormones thyréotropes, de croissance et lutéotropes étaient toujours présentes alors que l'ACTH avait disparu.

ZUSAMMENFASSUNG

Hypophyseneinpflanzung bei zerebraler Kinderlähmung.

Einpflanzung von aufbewahrten Ochsenhypophysen ist bei 84 Kindern ausgeführt worden. 65 von ihnen litten an zerebraler Kinderlähmung und 19 an verwandten Krankheiten. Verbesserung wurde in 19 Fällen (22,5 prozent), leichte Verbesserung in 45 Fällen (53,5 prozent) beobachtet. Die Bestimmung des Hormongehalts hat gezeigt, dass das TTH, das Wachstumshormon und das LTH sogar nach einer Aufbewahrungszeit von einem Monat noch vorhanden waren, während das ACTH verschwunden war.

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Hydrocephalus and Disability

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The prognosis in hydrocephalus is not nearly so grave as used to be thought, for a considerable proportion of these patients are now growing up into useful citizens. Their physical and mental handicap is not so much a result of actual thinning of the cortex as of associated malformations and secondary changes. The importance of the inter-relationship between mental and physical ability cannot be over-emphasised.

THERE is a general belief that few children with hydrocephalus survive, and that most of the unfortunates who do are so severely handicapped that they have to be cared for in an institution. A recent investigation (Laurence 1958) showed that many do survive and without surgical intervention, the disease being arrested spontaneously. Out of 81 survivors, 25 were physically and mentally normal, but some of the others were severely handicapped. It is hoped here to pursue some of the causes of this handicap, which in many instances is due not to the hydrocephalus itself but to the primary condition which gave rise to that hydrocephalus.

Before examining the causes, it may be useful to give a brief account of the circulation of the cerebrospinal fluid and the pathology of hydrocephalus.

Cerebrospinal Fluid Circulation

Physiologists believe that the cerebrospinal fluid is formed by the choroid plexus, but cannot agree as to the amount produced, though this is probably no more than 100 ml. per day. The plexi are frond-like structures, one situated in each of the lateral ventricles and extending through the foramina of Monro into the third ventricle, and a third plexus is found in the fourth ventricle. Fluid passes from the lateral ventricles, through the wide foramina of Monro into the third ventricle. From there it flows through the narrow aqueduct of Sylvius into the fourth ventricle. This aqueduct, though variable, is about 1 cm. long and has a diameter often no more than 0.1 cm. The cerebrospinal fluid then leaves the ventricular system by three exit foramina and flows into the basal cisterns and from there into the general subarachnoid space. In doing so it has to pass the narrow tentorial opening before reaching the large cerebral subarachnoid area and arachnoid villi. The fluid seems to be absorbed into the blood-vessels of the whole pia and arachnoid membranes. The classical experiments of Dandy and Blackfan (1914) suggest that four-fifths of the fluid is absorbed from the subarachnoid space above the tentorium and the remaining fifth from the posterior fossa and spinal subarachnoid space.

The Pathology of Hydrocephalus

Hydrocephalus is best defined as the excessive accumulation of cerebrospinal fluid within the cranial cavity. Certain forms of cerebral atrophy may be associated with such an accumulation of fluid, but this is only secondary to the loss of brain substances—i.e., hydrocephalus '*e vacuo*'—and will therefore not be discussed further.

Theoretically, hydrocephalus may be caused in three possible ways (Table I).

TABLE I—CAUSES OF HYDROCEPHALUS

Over-production of C.S.F.	Choroid plexus papilloma and villous hypertrophy.
Obstruction to C.S.F. pathway	Within ventricular system: Foramina of Monro; Aqueduct; Exit foramina. Subarachnoid space: Arnold-Chiari malformation; Basal cistern block. Outside C.N.S.: Skull abnormalities
Defective absorption of C.S.F.	? ? Venous sinus thrombus.

By far the most important is that resulting from obstructive lesions—among others, malformations and inflammations.

Malformations

The most frequent malformation associated with hydrocephalus is spina bifida cystica, the anatomy of which is particularly relevant in connection with the physical disabilities.

In the common spina bifida occulta, the mildest abnormality, the spines have failed to fuse, but the meninges and the spinal cord remain within the spinal canal. Although some cases show spinal-cord abnormalities, which may lead to leg deformity and weakness or urinary troubles, these cases are only very rarely associated with hydrocephalus.

The true meningocoele, not very often seen, is a lesion where, in addition to the spina bifida, the meninges but not the cord leave the spinal canal. Cord abnormalities are common (Cameron 1956), and account for the associated spinal palsies. Brain abnormalities, such as malformations of the aqueduct and more rarely the Arnold-Chiari malformation, account for few cases of hydrocephalus.

Myelocoeles and meningomyelocoeles are the commonest and are basically the same lesion, since they have a common embryological basis. Early in intrauterine life the neuro-ectoderm forming the neural plate fails to close over to form the neural tube, and the spinal-cord tissue remains on the surface. This is seen at birth as a raw and apparently ulcerated central area, usually surrounded by a blue and semi-transparent membrane which represents the meninges, and through which nerve-roots and blood-vessels may be seen. This defect may epithelialize early in neonatal life, but it more usually becomes infected, with unfortunate results, as will be discussed later. In addition to these abnormalities of the spinal cord, which usually produce more or less severe motor and sphincter paralysis, many cases show other cord malformations such as diastomatomyelia, hydromyelia and syringomyelia. Nearly all cases are associated with some degree of the Arnold-Chiari malformation and many cases have aqueduct malformations.

Two aqueduct malformations may be found in combination with spina bifida cystica—forking and simple stenosis (Fig. 1). In forking the aqueduct breaks up into two or more channels, one of which usually communicates between the third and fourth ventricles. In simple stenosis, on the other hand, the aqueduct may retain its normal outline but is markedly reduced in diameter. In both forms there is no excess of gliosis and the ependymal lining is intact.

Some form of Arnold-Chiari malformation is found in nearly all cases of myelocoele (Fig. 2). This is an anomaly of the hind-brain, where the cerebellum, often poorly lobated, has a posterior midline prolongation of vermis for a variable distance down the spinal canal. The medulla too is malformed and elongated and

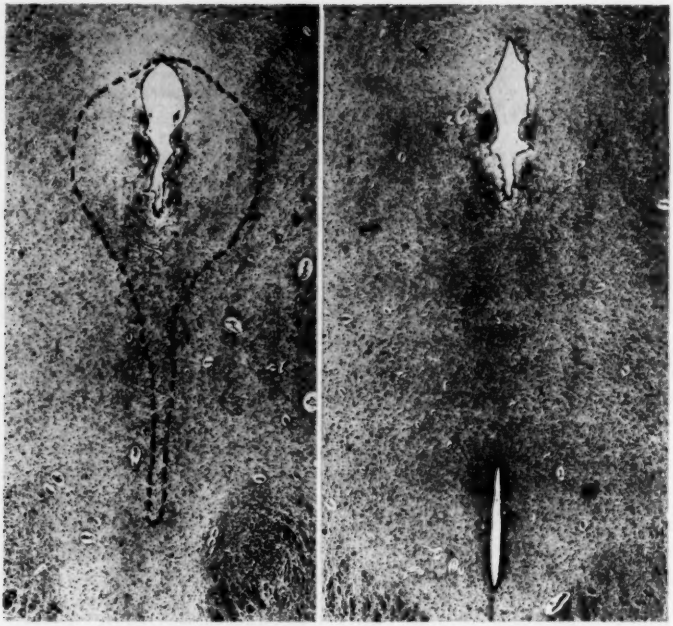

Fig. 1. Photomicrographs of simple stenosis (left) and forking (right) of the aqueduct of Sylvius. The outline of a normal aqueduct is drawn in in black. (Haematoxylin and eosin x 16)

extends below the level of the foramen magnum. Lying between the cerebellum and medulla is a narrow elongated fourth ventricle, with the choroid plexus often forming a compact mass near the tip of the cerebellum prolongation. The whole malformation is generally covered by thickened congested meninges. The congestion may in time lead to thickening and fibrosis and to complete obliteration of the basal cisterns or to occlusion of the fourth ventricle exit foramina.

Besides these two common malformations in the aqueduct of the hind-brain, other brain abnormalities which in themselves do not give rise to hydrocephalus are frequently found in cases of myelocoele. Among these are microcephaly, microgyration, ectopic subependymal grey matter, and abnormal thalamic nuclei.

The aqueduct anomalies and Arnold-Chiari malformations may in themselves lead to hydrocephalus, through the congestion and eventual fibrosis of the posterior fossa meninges and overcrowding of structures in the upper cervical spinal canal and in the small posterior fossa. More important in the production of hydrocephalus in these cases is the effect of ascending meningitis from the raw unepithelialized spina bifida sac, which leads to postinflammatory obstructions, superimposed on any congenital blocks already present. Thus, in addition, blockage of the foramina of Monro, the exit foramina, the aqueduct, or the basal cisterns may be found in most of these cases (Laurence 1959 *b*).

Inflammations

Inflammations not superimposed on central nervous system malformations account for a large number of cases of hydrocephalus (*vide* Table II). The foremost of these—bleeding into the C.S.F. pathway—most commonly results from birth trauma, especially in premature and

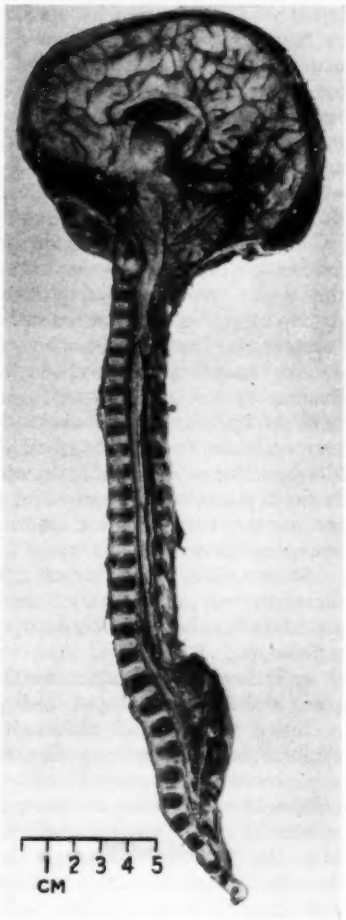

Fig. 2. A hemisected skull and spinal column, showing the Arnold-Chiari malformation. The elongated medulla is not only largely intraspinal, but is also kinked upon the cord. A well-developed cerebellar tongue is shown. The myelocoele in the lumbo-sacral region is typical, with nerve roots leaving the neural plate to gain the root canals. The cord shows slight hydromyelia.

first-born infants. Recent investigations indicate that such trauma may occur in normal deliveries, often without any signs suggestive of intracranial mischief. It is important to remember that when trauma sufficient to produce C.S.F. bleeding or a subdural haematoma has been inflicted, parenchymal damage, in the form of minute diffuse lesions, mistakenly called 'congenital encephalitis' by some older authors, or larger intracerebral haemorrhages or even softenings are produced at the same time. It is these associated lesions that account for at least some of the disability often seen in cases of hydrocephalus of this causation. Other causes of bleeding into the C.S.F. leading to hydrocephalus, especially in children, are haemorrhage from berry aneurisms, operations or head injuries, where extensive bleeding into the basal cisterns has taken place (Foltz and Ward 1956).

Infections which lead to postinflammatory blockages are usually bacterial, but may sometimes be viral or even toxoplasmal in origin. Such a meningitis may be mild and remain undetected, and even a severe *Bact. coli* infection in the neonatal period may be detected only by chance after a diligent house-physician has performed a routine lumbar puncture on an

TABLE II—AETIOLOGY OF HYDROCEPHALUS (100 Post-mortem examinations)

Malformation:		46
Alone	14	
With infection	30	
With trauma	2	
Inflammation:		50
Infection alone	17	
Trauma alone	22	
Unknown (probably infection or trauma)	11	
Tumours		4

infant who is ailing but has no symptoms to suggest a meningitis. The mild and the neonatal cases of meningitis tend to have the severest effects because they may remain untreated for a long time. It must be emphasised that a meningitis is usually associated with an encephalitis and perhaps with extensive thrombosis, which accounts for some of the associated disabilities.

The severest result of such inflammatory lesions is an organised pyencephalus, where the whole ventricular system becomes converted into a series of intercommunicating cysts. Less dramatic consequences are the denuding of the ependyma after damage by the inflammatory agent. For once this ependyma is denuded it does not regenerate and the subependymal glia proliferates. This process is of little importance in most places in the ventricular system, but in the narrow aqueduct, and very occasionally in the much wider foramina of Monro, this subependymal glial proliferation may occlude the lumen. The aqueduct can also be blocked by pus or by a blood-clot.

An inflammatory reaction usually takes place also in the meninges, leading to an occlusive fibrosis and blockage of the cerebrospinal-fluid pathway. Generally this produces a basal-cistern block, with the arachnoid bound down at the edges of the cisterns (Fig. 3). In some instances, however, the fibrosis involves the meninges near the exit foramina, thus resulting in an exit-foramina block.

In many cases a combination of post-inflammatory lesions are present and an aqueduct stenosis may be associated with an exit foramina block or a basal cistern block.

Fig. 3. Hemisected skull of a case of basal cistern block. The dilated basal cisterns, outlined by indian ink injected post-mortem, are greatly dilated, at the expense of the cerebellum behind, and the floor of the third ventricle and the pituitary stalk anteriorly. The dilated lateral ventricle can be seen through the foramen of Monro and the fenestrated septum pellucidum.

The Natural History

Although hydrocephalus is a very serious and often fatal disease, some patients do survive without treatment after natural arrest of the condition. Many authors note this natural arrest but do not give any idea of its frequency. Laurence (1958) carried out a follow-up investigation of a large series of untreated and unselected cases of hydrocephalus, seen by one neurosurgeon over a 20-year period, in order to investigate this particular problem. The results were most unexpected. Of 182 patients, 81 were found alive and well with hydrocephalus arrested spontaneously, 3 others were probably well but untraceable, and the remainder had either died or the hydrocephalus was still not quiescent (Table III).

The intellectual performance of the survivors with arrest, and their physical condition, are both of the greatest importance, for on these factors depends their future place in the community. Many factors are responsible for this handicap.

Physical Disability

Severe hydrocephalus may in itself be responsible for physical disability, though a less severe form, especially after natural arrest has taken place, need not be associated with any abnormalities at all. Spasticity and inco-ordination, almost certainly caused through stretching of

TABLE III—NATURAL HISTORY OF HYDROCEPHALUS (182 patients)

	No. of Cases	
Alive:		
Hydrocephalus arrested	81	(46.2%)
Untraced (presumed alive)	3	
Still progressive	9	(5.0%)
Dead	89	(48.8%)

motor tracts and compression of nuclei by the dilating ventricular system, is very commonly seen. Stretching and compression of cranial nerves themselves frequently cause ocular palsies, though they are often transient. Vision is not usually completely lost in spite of mistaken views to the contrary set out in most textbooks. This misconception probably arose because of the temporary lack of response to stmuli in progressive hydrocephalus, found together with pale discs, which is normal in infants and is mistaken for primary optic atrophy.

The blood-supply may become compromised by the high cerebrospinal-fluid pressure. This and thromboses of blood-vessels and sinuses may lead to infarctions of the cerebrum, and thus to motor palsies. Smaller anoxic areas give rise to cortical scars. Even if these do not produce palsies, they may act as epileptogenic foci, though focal discharges are not often found on the electroencephalogram of hydrocephalic infants. Only 16 of 50 such infants examined at The Hospital for Sick Children, London, showed discharges, and only 3 of these infants with discharges actually had epileptic fits (Laurence and Pampiglione 1960).

Much of the physical disability of hydrocephalic infants is due to associated malformations, or due to the original lesion that led to the development of hydrocephalus. For example, the physical disability not due to the hydrocephalus itself, in postinflammatory hydrocephalus, may have been produced by the encephalitis or other parenchymal damage which accompanied the inflammation. Meningeal fibrosis, which itself gives rise to the cerebrospinal-fluid blockage, may at the same time involve blood-vessels and nerves. On the other hand, in cases associated with myelocoeles, physical disability may be due to three distinct causes—the myelocoele itself, associated malformations in the central nervous system or elsewhere, and superimposed infection.

The myelocoele is responsible for very severe neurological effects in many cases. The pattern of these neurological lesions is to a large extent dependent on the position of the myelocoele (Laurence 1959*a*). High spina bifida cystica leads predominantly to motor loss, perhaps affecting mainly the trunk and thighs, while sphincter paralyses are unusual. In lesions lower down the spinal axis, paralyses of the legs are generally associated with severe sphincter involvement. Cases of myelocoele in the sacral region, on the other hand, may have sphincter involvement only, the legs being almost completely spared (Fig. 4). Sensory loss, too, varies according to the site of the spinal lesion.

Consequent on the motor deficits, various other troubles ensue. For example, muscle inbalance and paralyses lead to talipes equinovarus, dislocation of the hips, and contractures or scoliosis. Sphincter paralyses lead to chronic urinary infection or chronic constipation, with all their consequent evils and sensory loss, to deep trophic ulcers, and perhaps to osteomyelitis of the underlying bone.

Other congenital malformations are common, both within the central nervous system and elsewhere. Renal tract malformations, such as congenital hydronephrosis and double ureters, which also predispose to renal-tract infections, and skeletal anomalies are most frequently found. Severe kyphoscoliosis, resulting in respiratory embarrassment, may follow abnormalities of the vertebrae which are often combined with rib anomalies. Persistence of the neurenteric canal or its

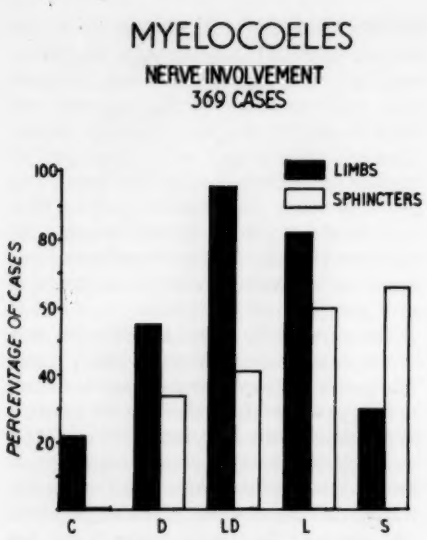

Fig. 4. The 369 cases of myelocoele studied were divided up according to the site of the lesion. The percentage of the cases in each group showing motor (in black) and sphincter involvement (in white) is represented by the length of the column.

The letters represent the position of the lesion. C = cranial and cervical; D = dorsal; L.D. = lumbodorsal; L = lumbar and lumbosacral; S = sacral only.

remnants is often found and may give rise to a variety of troubles, such as sacral dermoid, various cysts or pilonidal sinus. Spinal-cord malformations, such as diastematomyelia, with its bony spur, severe hydromyelia or progressive syringomyelia, regarded by some as congenital, may lead to progressive cord signs.

This may wrongly suggest that physical impairment is the rule, but of the 81 survivors with natural arrest, 27 (33 per cent) were normal physically. A further 15 cases did suffer from disability, but these were still able to get about unaided and in most cases were not compelled to attend special schools on account of their physical disability. There were 22 severely handicapped who always had to attend special schools, or even had to receive home schooling. Only 17 cases were completely bed-ridden and generally confined to institutions.

This series is not representative, since it contains fewer cases of spina bifida than are found in most series of hydrocephalic patients, but these figures do demonstrate that the outlook for survivors is not so gloomy as is usually suggested.

Mental Impairment

Intelligence assessments, mostly intelligence tests done by Stephen Coates, were obtained on all the 81 cases. Totally unexpected results were obtained (see Fig. 5 and Table IV). The average intelligence of the whole group was 69.7 per cent, with an approximately normal distribution, but a standard deviation about twice the normal. If the 17 worst cases, with I.Q.s below 40, are excluded, then the mean I.Q. of the remainder is 84, which approximates to the normal intelligence range.

The causes of this reduction of intelligence are threefold. Hydrocephalus itself undoubtedly leads to impairment of intelligence, but this is often surprisingly slight. In a comparison between the I.Q. and the thickness of the cerebral mantle estimated by air encephalography, the long-accepted maxim, that persons with a cortex less than 2 cm. thick are likely to be severely mentally handicapped, was shown to be incorrect. Two children with a cortex less than 0.5 cm. thick had I.Q.s of 85 and 100, and a considerable number of cases with the cortex only a little thicker had quite normal intellects, as judged by the usually accepted tests (see Fig. 6). A similar state of affairs exists between intelligence quo-

TABLE 4—INTELLIGENCE ASSESSMENTS (81 patients)

	No. of Cases	
Normal intelligence (I.Q. over 85)	33 (41%)	59 (73%)
Below normal intelligence, but educable (I.Q. 50–84) ..	26 (32%)	
Ineducable (I.Q. under 50)		22 (27%)

tients and the head circumference (see Fig. 7).

Although some of these patients score very well on the conventional intelligence tests they are not so adaptable as normal children of similar intelligence. The more severely impaired children score quite well on memory and particularly verbal tests, but not so well when they have to grapple with problems requiring reasoning.

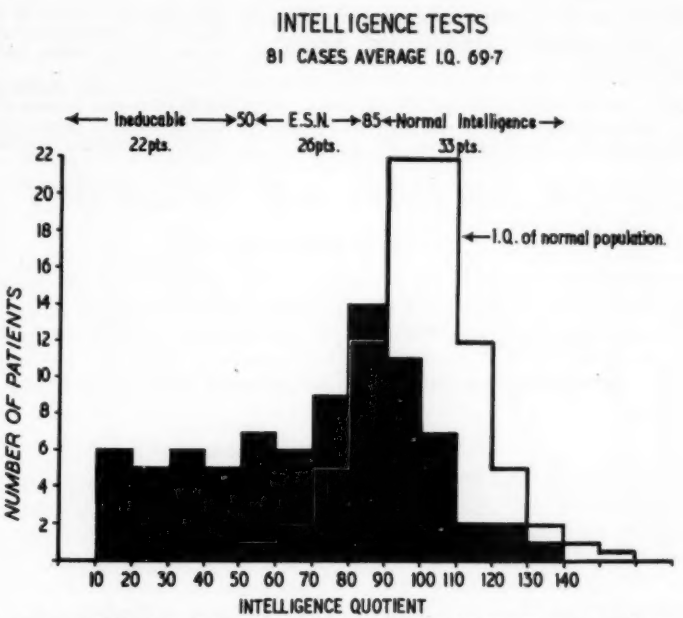

Fig. 5. Distribution of intelligence assessment results in 81 cases, with an average I.Q. of 69.7. The superimposed line represents the distribution of the same number of test results in a normal population.

A considerable amount of the intelligence deficit is due to damage done by the infection or trauma which led to the original hydrocephalus. Some of the most severely impaired children in the series have comparatively little hydrocephalus. Others have associated cerebral malformations which may account for some of the loss.

Finally, mental and physical performance are to a large extent mutually interdependent. A child with severe physical disability will score low on most intelligence tests, because most of these tests involve at least some physical skills. Severely handicapped children also have difficulty in communicating with others of the same age, either because they cannot play freely with others, being isolated at home or in an institution, or because they lack one or more of the five senses. Older children may require so much hospital treatment at the expense of their schooling that they do not receive the same educational opportunities as their normal contemporaries.

General Conclusion

Hydrocephalus is a serious condition, with many different aetiologies, but a much more optimistic attitude should be adopted to it. This is so not only because a larger proportion of these children survive than was realised but also because the survivors are not so severely handicapped as most clinicians thought. Many are growing up into useful citizens capable of earning their own livelihood.

On the other hand, the handicap of hydrocephalic patients is to a large extent due to the primary infection or injury or the associated malformations, rather than to the hydrocephalus itself. Dilatation of the ventricular system, with the usual concomitant enlargement of the head and reduction in the thickness of the cerebral

mantle, seems to have only a lesser effect on the patient's physical and mental performance.

Achnowledgments: I am indebted to the Editors of the *Annals of the Royal College of Surgeons*, of the *Quarterly Review of Paediatrics* and of the *Lancet*, for permission to reproduce Figs. 2, 3 and 5, respectively, which were previously published in their journals.

Thanks are also due to the Research Committee of The Hospital for Sick Children, Great Ormond Street, London, under whose aegis most of the work on which this paper is based was carried out.

CORTICAL THICKNESS AND INTELLIGENCE

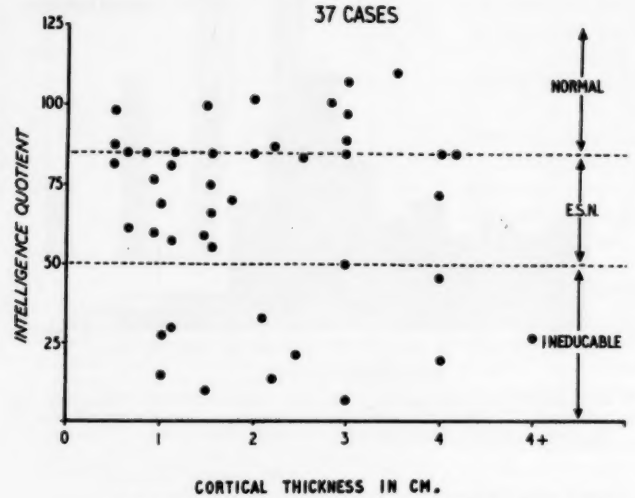

Fig. 6. Distribution of intelligence assessment results when plotted against the thickness of the cerebral mantle in the 37 patients in whom reliable encephalographs were available.

HEAD SIZE AND INTELLIGENCE

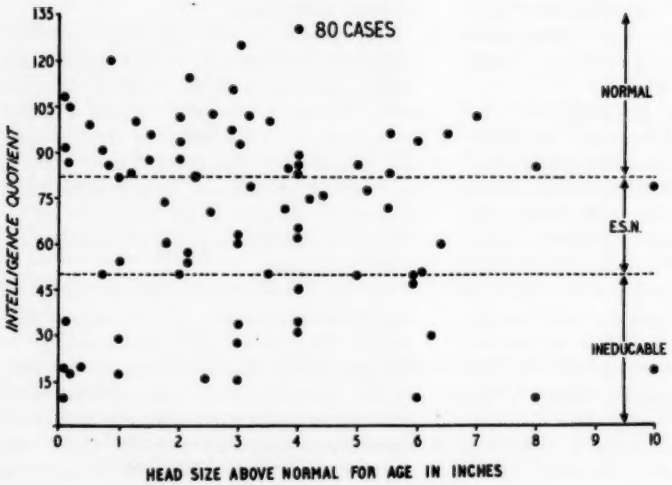

Fig. 7. Distribution of intelligence assessment results when plotted against the head measurement, measured in inches above the normal for the age and sex.

SUMMARY

A brief account is given of the cerebrospinal fluid circulation, and the pathology of hydrocephalus and associated conditions, together with the natural history of untreated hydrocephalus. The causes of the physical and mental handicap of patients with hydrocephalus are discussed and it is concluded that much of this was due not so much to the hydrocephalus as to the associated lesions. It is suggested that the outlook in these cases should be much more optimistic.

RÉSUMÉ

Bref exposé de la circulation du liquide céphalorachidien, des lésions de l'hydrocéphalie et des états connexes ainsi que de l'évolution spontanée de l'hydrocéphalie non traitée. Les causes de diminution physique et mentale des malades atteints d'hydrocéphalie sont discutées et on conclut que la plupart de celles-ci sont dues moins à l'hydrocéphalie qu'aux lésions associées. L'auteur pense qu'un pronostic plus optimiste devrait être porté.

ZUSAMMENFASSUNG

Die Zirkulation der zerebrospinalen Flüssigkeit und die Schädigungen bei Hydrocephalus und verbundenen Zuständen sowie der spontane Verlauf des unbehandelten Hydrocephalus werden kurz dargestellt. Die Ursachen der körperlichen und geistigen Beeinträchtigung der Kranken mit Hydrocephalus werden erörtert und es wird daraus geschlossen, dass man die meisten von ihnen weniger dem Hydrocephalus als den verbundenen Schädigungen zuschreiben muss. Der Autor meint, dass die Prognose optimistischer sein sollte.

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A Visit to Paris

A GROUP of six paediatricians and registrars, led by Dr. Ronald Mac Keith, spent a week in Paris in March under the auspices of the National Spastics Society, to study some aspects of neurology in the newborn infant at the Clinique Baudelocque. This is a big maternity hospital in the heart of Montparnasse, standing in the grounds of and still incorporating some of the buildings of the ancient convent of Port-Royal, famous for Pascal and the Jansenists. It contains a Neonatal Research Unit under Dr. Alexander Minkowski, whose medical and nursing staff look after the newborn and premature infants. The unit is engaged in many forms of research, and one of their chief interests is the neurology of the neonate.

Mme. Saint-Anne Dargassies and newborn friend.

This interest was first awakened by Dr. André-Thomas, who some 15 years ago turned his attention from adult neurology to that of newborn infants, and his work is now carried on primarily by Mme. Saint-Anne Dargassies, who was indefatigable in demonstrating and imparting the French methods to her visitors.

The André–Thomas Method of Examination

The orthodox method of neurological examination, based on the anatomy of the nervous system and conducted with a view to localising a lesion, is of little value in the very young child. The neurological examination devised by André-Thomas and his school, based on the physiology of the nervous system, regards the newborn infant as a subcortical, tonic individual exhibiting definite automatisms, and reveals a great deal more information. The details of the examination are described by André-Thomas and Mme. Saint-Anne Dargassies in a supplement to this *Bulletin*, so that only a brief résumé need be given here.

The infant is carefully watched for his general behaviour, his cry and his movements. Muscle tone is carefully assessed and compared on the two sides, by palpating the muscles for their *consistency*, by testing their full range of movement at a joint to assess their *extensibility*, and by estimating the ease with which muscles allow the segment of a limb to be 'flapped' to and fro to assess their *passivity*. Finally, various automatic reactions and reflexes are elicited. The reactions are complex responses to specific stimuli, liable to variation and modification by the infant's physiological state, such as sleep, hunger, repletion or crying. The interpretation of any apparent abnormality, especially if it

is an isolated phenomenon, therefore requires care. The reactions are best sought when the infant is fully awake, ready for a feed but not crying, and for convenience's sake are best elicited on a segmental basis.

In the head and neck, the ocular signs examined include the doll's-eye reflex, contraction of the pupil to light, closure of the eyes to a strong light, and, after the first ten days, an attempt to follow a soft light such as that emanating from a window. Facial reactions are then elicited by tapping the forehead (McCarthy reflex), by firmly rubbing the auricle, and by gently stroking the angles of the mouth, the upper and the lower lips in turn (cardinal points or rooting reflex). In the limbs, the Moro and grasp reflexes are elicited, together with the effect of attempting to withdraw the finger. The grasp reflex of the toes is also noted and the crossed extension reflex of the lower limbs. The infant is held upright and the soles placed on the firm surface of the examination couch to observe definite extensor thrusting movements of the lower limbs downwards, and walking movements. In examining the trunk's reactions the infant is held in the air in ventral suspension, and the skin alongside the dorsal spine is stimulated to elicit flexion of the trunk towards the side of the stimulus. The baby is held firmly by the pelvis against the examiner's body with the trunk drooping forwards, firm pressure is exerted against the soles of his feet, and the infant's body then rights itself by extension of the trunk.

The premature infant, born at 28 weeks of gestation, presents a very different neurological pattern. He has a generalised marked hypotonia. In contrast to the infant born at full term, the arms are only slightly flexed and often extended. When he is supine his thighs lie on the bed in a frog-like position. A limb lifted and released will drop heavily without any rebound. At the same time, however, there are many and varied spontaneous movements. These may be localised minor contractions of isolated muscles or muscle groups, seen especially in the face, the hands and the feet. At other times they are slow, majestic, purposeless assymetrical flexions and extensions of limbs. Perhaps the most characteristic spontaneous movements are sudden, vigorous and repetitive explosive movements of the limbs and trunk. These are spasms or quivering trepidations of arms and legs, and rolling and writhing movements of the trunk and pelvis, leading sometimes to remarkable changes of posture and position in the incubator.

The infant born at 32 weeks will show most of the primary automatic reactions present at term, though they are often poorly marked. The righting reaction of the head on the neck is not present. The walking reaction is transitory, and when he does walk the child does not place the sole of his foot on the ground but walks on the tips of his toes.

In infants born at 36 weeks the neurological pattern is that of an infant born at term.

Value of the Neurological Examination

The neurological examination of infants is valuable in detecting the presence or degree of cerebral trauma, and it provides a systematic method of following the day-to-day progress of such children. It is also a useful adjuvant to other clinical signs in determining the degree of immaturity in small babies or babies born before the expected day of delivery, and it is helpful in charting their gradual maturation. The neurological examination does not, however, help in localising lesions, since it is difficult to say which alterations in tone or reactions have a consistently pathological significance. Nor does it help in prognosis, for at the present state of knowledge no inference can be drawn from any particular abnormality as to the infant's future development.

All infants born in the Clinique Baudeloque who have been premature or have shown neurological abnormalities are followed up at the Clinique. In the older child, too, the central nervous system is studied largely from a physiological, psy-

chological and psychomotor point of view. At these follow-up clinics, in addition to an ordinary neurological examination, careful note is taken of muscle power and tone and the projection of the body image in space. This is tested mainly by making the infant imitate gestures and stances, at first simple and later quite complex. Anticipation of such movements, too frequent repetitions, or unduly long persistence in attitudes, help to shed light on the child's psychological disposition. The mother is also questioned about the child's emotional state, his attitude to work and play and his sleep, and finally the children are seen by a child psychiatrist.

Other Activities

Electroencephalograms are frequently performed on the infants under the care of the service. This is purely a research project, and a large number of records of premature infants, brain-damaged infants, and normal infants have now been obtained. As yet, however, the help given to the clinician is limited, and no guide to prognosis is possible. Nevertheless, valuable information is being collected on normal and abnormal wave patterns at all ages and stages of early life.

The many other topics discussed during our stay at greater or lesser length included jaundice in the premature infant, the oxygen-carrying power of foetal haemoglobin, hyaline membrane disease, cross infection with special reference to neonatal meningitis caused by *Esch. coli*, and many others.

One afternoon the group visited the Centre International de l'Enfance, to be welcomed by the Director, Dr. Etienne Berthet, and Dr. Nathalie Masse. The Centre is in a château in the Bois de Boulogne and is concerned with the general distribution on an international level of information on all aspects of paediatrics. This is done by means of courses and seminaries for doctors, midwives, nurses and social and welfare workers. Medical biological and medical social research is carried on, not only in France but also in other countries, and well-equipped virus research laboratories are run by the centre. A big reference library is available and for a small fee any doctor in any country can be sent a monthly list of references on subjects of his choice. The centre also publishes the monthly review, *Courrier*, which presents general surveys of topical paediatric problems, as well as selecting and abstracting the important articles on paediatrics in all its aspects published in the international journals.

The group also visited the Hôpital des Enfants Malades for a clinical meeting presided over by Prof. Robert Debré, where some extremely interesting cases were presented and discussed in detail by the French paediatricians.

We left Paris greatly impressed, not only by what we had seen and learnt and the original lines of thought and pioneering spirit of research displayed by all the members of the neonatal unit, but also by their individual kindness, helpful friendliness, and true Parisian gaiety and charm.

LEONARD HAAS

EDITORIALS

The First Breath

FEW events in Nature inspire more wonderment than the first breath of the newborn child and few are more difficult to investigate scientifically. It is a moment of deep import. The onset of pulmonary respiration means to the mother and her attendants that the baby has survived the process of gestation and birth and has taken a critical step towards adaptation to extra-uterine life. To the baby just now deprived of his mother's oxygen, and so in momentary extremity of asphyxial death, there comes a renewal of the vital element, and life continues.

Failure to breathe at birth almost always means that the baby is very immature, or malformed, or has suffered some harm during parturition. Resuscitation may restore life, but in those that survive there is a possibility that permanent, if non-progressive, damage has been done to the brain.

Preparation

During foetal life the lungs are not used for breathing and little blood flows through them. The oxygen requirements of the foetus are met by the blood traversing the placenta and returning to the foetus via the umbilical vein. As pregnancy advances the foetal lungs change their structure and become better adapted to the function of breathing. This maturation is important. Essentially the lungs are composed of millions of small ballons (alveoli) which all open into the major air-passages. The walls of the alveoli are permeable to gases such as carbon dioxide and oxygen. The former diffuses very readily across, oxygen rather less so. In the spaces between the alveolar walls, blood-vessels deliver up the carbon dioxide elaborated by the metabolism of the body and take up the oxygen contained in the air drawn into and filling the alveoli.

The changes in the lungs before birth are a progressive thinning of the alveolar walls to permit easy diffusion of gases and an increasing vascularisation of the lungs. Until the first breath or breaths fill the alveoli with air they are collapsed or airless—in a condition of atelectasis. As the air-sacs become filled the lungs expand and will not again collapse unless the inflow of air is obstructed.

Apparatus

In health, pulmonary respiration is so controlled as to require the least expenditure of energy. The volume of air breathed increases to meet greater needs due, for instance, to exercise or rarified atmosphere, but is reduced in sleep. This control is a function of a specialised part of the brain, the 'respiratory centre'. This receives information on the body's chemical state and on whether it has too much or too little carbon dioxide and oxygen, both directly from its own blood supply and notably about oxygen lack— also by relay from peripheral 'chemoreceptors' situated near the carotid arteries.

The respiratory centre also receives afferent impulses from the lungs them-

selves, indicating their state of expansion or distension. This is important for maintaining the normal rhythm of respiration. Changes in the rate, rhythm or volume of respiration are dictated by the respiratory centre and transmitted to the muscles which work the respiratory 'pump'. These muscles, and especially the diaphragm and intercostal muscles, bear the responsibility for altering the capacity of the bony thorax. Increasing the capacity induces a negative intrathoracic pressure and air is drawn into the lungs (inspiration). Decreasing the capacity allows the lungs to relax and expels the air (expiration).

Expansion

Without these essential mechanisms the newborn infant cannot breathe, and initially he has to accomplish yet another act. This is to convert his collapsed, airless alveoli into inflated air-sacs and at the same time divert into his pulmonary circulation a large volume of blood which has hitherto by-passed the lungs. The first is a mechanical feat, because the collapsed alveolar walls are stuck together by the surface-tension effect of a thin layer of fluid. To open them up demands a very great negative pressure. Assistance comes in two forms. Primarily, during the passage through the birth canal, the baby's chest has been tightly squeezed, ridding the air-passages of excess fluid, and the recoil of the chest probably helps expand the lungs. Co-operatively, the establishment of the pulmonary circulation, which is part of the complicated rearrangement of circulation in neonatal life, actively helps the opening of the alveoli. Consequently the first breaths involve more exertion than subsequent ones and raise a query why all the air is not expelled from the lungs at each expiration. This would be disadvantageous; as would be an unequal filling of the alveoli resulting in progressive over-filling of expanded alveoli and non-filling of those in the collapsed condition. Both hazards are minimised by a surface-acting agent with 'detergent' properties, probably a muco-protein, present in the lining of the air-passages and alveoli and effective in reducing surface tension.

Investigation

The apparatus that the baby needs in order to breathe is highly developed during the latter weeks of foetal life and at full term is ready for pulmonary ventilation. In fact, the classic researches of Sir Joseph Barcroft suggested that the foetus near term was only prevented from making respiratory efforts by an active inhibitory process. It has now been shown that infants subject to anoxic stress *in utero* do make respiratory efforts which are, of course, functionless and may be harmful. This suggested that anoxia might be the most effective stimulus to respiration, acting possibly by removal of inhibition. The idea that the infant *in utero* became progressively less well oxygenated before term lent support to the hypothesis.

Dr. Stanley James (1960) has severely criticised the concept of intrauterine oxygen deficiency near term and has suggested that the oxygen deprivation shown by many infants at birth is an acute and transient result of disturbances during delivery. If this is correct, the idea that the foetus becomes gradually and progressively 'primed' for the onset of pulmonary ventilation requires revision; it does not destroy the view that anoxia is a potent stimulus for breathing.

As soon as the infant is divorced from his mother's oxygen supply he is in a state of oxygen lack until he starts to breathe air. At the same time there is

retention of carbon dioxide. The tendency to acidosis is further increased by accumulation of acids derived from the incomplete metabolism of glucose. The effect of anoxia and excess acidity (low pH) on cells is to cause a leak of potassium ions which emerge into the blood. The anoxic infant will therefore rapidly get profound and serious disturbances of acid/base and electrolyte balance. These biochemical changes initially stimulate respiration but if not corrected will later interfere with metabolic processes and cause the death of cells. The depressant effects will be felt by the cells whose metabolic needs are greatest—outstandingly, those brain cells with the greatest number of integrative and co-ordinating functions.

Barcroft observed two major patterns for the onset of respiration. The first a smooth, rapid onset of almost normal breathing which starts as a response to cutaneous stimuli; the other a gasping, inco-ordinate breathing found in depressed infants. Later work has filled in many of the details and confirmed the essential truth of these observations.

Initiation

In natural, normal childbirth the infant may be conditioned to brief periods of hypoxia during the regular, effective uterine contractions. These will not last long enough to depress the child's brain cells, and since there will be no potent analgesics and anaesthetics given to relieve the mother's discomfort, the infant's brain cells will not be saturated with potentially depressant drugs.

As the baby is compressed while traversing the birth canal the indwelling fluid is expelled from the respiratory and alimentary passages. At birth the airway is clear, the respiratory centre alert and responsive, and there is no accumulation of the metabolic by-products of anoxia. When the head emerges, an inspiratory effort takes place which is powerful enough to draw air into the air-passages and inflate the lungs. Simultaneously haemodynamic changes occur which establish the pulmonary circulation. These have been beautifully depicted by Barclay, Franklin and Prichard (1948) and since then by Prof. John Lind of Stockholm.

Maintenance

For many years it was thought that the lungs expanded rather slowly after the onset of breathing. However it is now known that the lungs are rapidly aerated with the first few breaths in normal infants. The most striking evidence for this are the X-ray studies of the Swedish workers led by Lind (Fawcitt, Lind and Wegelius 1960). Supporting evidence comes from physiological studies of the mechanics of respiration which show that lung function reaches normal values quite soon after birth. A continuing series of studies by Prof. Clement A. Smith and his group at the Boston Lying-in Hospital and by P. Karlberg in Stockholm have provided much information on this subject. It has been known for some time that the oxygen saturation of the blood in newborn infants reaches normal values within an hour of birth and this would be unlikely if large areas of lung were unexpanded.

A small, but by no means trivial, question is how does the infant direct the air-flow into the respiratory passages? The ingenious radiological studies of Bosma (1960) indicate that in some cases air is 'swallowed' into the pharynx and then passes into the trachea. This presents an interesting analogy with the 'frog-breathing' or glosso-pharyngeal respiration practised by some polio-

myelitis patients with paralysed respiratory muscles.

The scientific investigation of the first breath has revealed the extraordinary precision of nature's means of ensuring a smooth transition from intrauterine life to an independent existence. It has pointed out the multiplicity of factors involved and the inter-relationships that exist between the many body systems concerned. Most important, it has made us wary of interfering with natural events, and if interference is inevitable, or if pathological processes intervene, we can now step in more wisely and with greater knowledge.

THOMAS OPPÉ

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Right and Left*

At a time when many vast psychological problems—race prejudice, clashes of basic political beliefs, mental deficiency, sexual deviations, social delinquency, the rearing of children, or the growing pains of new, born nations—loom large, the difficulties of telling the right hand from the left, or one finger from another may seem trivial, and a book-long discussion of them a demonstration of ostentatious expenditure of psychologist man-hours. It is clear, however, that these simple faculties are related to wider issues—to biological survival, for instance, in choosing which side of the road to ride on or to walk on, this side of the English Channel or the other; or of social survival, knowing which hand predominantly to hold your fork in, this side of the Atlantic or the other. Further afield they link up with such interesting neuropsychological matters as dominance of hand or of cerebral hemisphere; such fashionable concepts as the schema or image of the body; and such far-reaching symbolisms as that involved in sitting on the right hand of God, or not letting your left hand know what your right hand does.

Perhaps because I have never achieved confident fluency in the use of right-left concepts myself, and only too often feel my fingers to be all thumbs, I have had the stubbornest difficulties in following the literature on these topics, and have hitherto been less than grateful to Dr. Benton and his co-workers for adding several papers to it. From this intellectual-emotional block Dr. Benton's book has now released me. It gives a critical analysis of his own and others' work on the normative aspects of right-left discrimination and of finger localisation and of their disturbances both developmental and acquired. It gives also a very handy summary of Dr. Benton's well-worked-out series of tests. Methodologically these are of special interest in the careful differentiation of the nature of the tasks involved. They range from the obeying (with the eyes open or blindfold) of a simple verbal command ('show the left hand'), through a double uncrossed command ('touch the left eye with the left hand'), to a double crossed command ('touch the right eye with the left hand'); and to commands to do the same with reference to a person facing one (which requires an understanding of how right and left shift with rotation of the body through 180 degrees) and to name as right or left a part touched by the examiner (which requires the ability to respond with a verbal symbol).

It is not surprising that, having designed and used so discriminating an instrument, Dr. Benton discusses critically the status of some of the peculiar associations of behavioural disorders which have been reported and whose implications for notions of brain-behaviour relationships are important. Subjected to analytical attack, the association between body schema and language and number skills begins to look adventitious. It is well known that disorders of right-left discrimination, of speech, and of disease of the dominant hemisphere may be associated. Dr. Benton's work indicates that the association of these disorders may be a reflection of the functional dependence of the more complex kinds of right-left discrimination on language, the basic ability to differentiate between right and left sides of one's body being not so dependent and having no special relation-

* Right-left Discrimination and Finger Localization. By Arthur L. Benton. *New York*: Paul B. Hoeber, 1959, 181 pp., £2 10s.

ship with the dominant hemisphere. Dr. Benton and his colleagues failed to confirm the supposed association of disorders of finger discrimination and of arithmetic. Dr. Benton's probings extend to the Gerstmann syndrome, that bizarre tetrad of disorders—right-left discrimination, finger discrimination, writing and arithmetic —thought to be caused by focal disease of the parieto-occipital region of the dominant hemisphere. Is it, he asks, a syndrome in the sense that its four components have a stronger association with each other than with other behavioural deficits, or is it only one of many equipotential combinations of deficits to be found in disease of the dominant hemisphere? Clinical records, he says, are inadequate to settle this kind of problem, for the methods used to test the disorders have been too diverse. It is essential to have neuro-pathological investigations matched with fully adequate behavioural studies of function.

What you discover depends on the tools you use. Dr. Benton demonstrates a precise set that we can borrow or adapt for further purposes, whether we are primarily psychologists, neurologists, or paediatricians. He has certainly given a heartening example of how to start to find what goes with what in behaviour disorders.

M. L. JOHNSON ABERCROMBIE

Prognostic Value of the Neurological Examination of the Newborn

in Relation to Later Neurological and Behaviour Disorder

DR. HEINZ PRECHTL, of the University of Groningen (Netherlands), recently gave a lecture[1] in connection with the Collaborative Project on Cerebral Palsy, Epilepsy and other Disorders of the Nervous System, now under way in a large number of centres in the United States, under the auspices of the National Institutes of Nervous Diseases and Blindness of the National Institute of Health. In recent years, he and his colleagues have studied the neurological findings in newborn infants, comparing infants with a relatively normal birth with those whose birth has been in some way complicated. They have now analysed the results of the neurological examination done at birth and the child's development as revealed by follow-up examinations after 2–4 years. The neurological examination made at birth attempts to be quantitative, analysing various behaviour and motor patterns.

Among 218 full-term newborn infants with a history of complications in labour, 42 per cent were classified on the neurological examination as non-pathological. The remaining 126 infants showed signs and symptoms, such as absence of certain reflexes, hyper-irritability or epilepsy. The highest proportion of neurological abnormalities was found in infants with a history of foetal distress and prolonged labour. There was a higher proportion of pathological findings in boys and in babies whose birth-weight had been under 6½ lb., although asphyxia was more common in larger babies.

A follow-up examination of the children was done after 2–4 years. Of the children who had shown abnormalities at their neurological examination at birth, 68 per cent showed neurological defects at the follow-up examination, compared with 8 per cent of the controls. Of the children who showed evidence of 'cerebral irritation' at birth in the series published by Craig[2], only 1 in 12 later had cerebral palsy or severe intellectual handicap. It is therefore important, in every case, to describe the degrees of abnormality found in the neurological examination. (It must be noted—whether or not it affects the result—that among those seen by Prechtl at follow-up examination there were 119 children whose birth examination had been abnormal, whereas only 18 children of the normal-birth control group were seen at the follow-up. Furthermore, 8 per cent of 18 children is 1½ child; this arithmetical curiosity makes one wish to see Prechtl's paper in closer detail.)

Prechtl made other observations

1. *Collaborative Project Reporter*, March 1960.

2. Craig, W. S. 'Intracranial irritation in the newborn: immediate and long-term prognosis'. *Arch. Dis. Childh.* 1950, 25, 325.

which may prove to be of very great importance. Neurological abnormalities detected at birth seem to have a prognostic significance in regard to the later development of behaviour problems. While abnormal findings at birth were in his series correlated with the emergence of behaviour problems later, the incidence of such problems was *not* correlated with the existence of unfavourable social surroundings. This suggests that, in this series, many behaviour disturbances were due to organic brain damage, and this may be true of other changes elsewhere—a most intriguing conclusion.

R. C. MAC KEITH

Infantile Spasms

ALTHOUGH first described over a century ago[10] the variety of infantile epilepsy variously known as infantile spasms, salaam spasms, lightning spasms, *tic de salaam* or *Blitz-Nick-Salaam-Krämpfe* has attracted less attention in Britain than in North America or Europe. Yet this condition is a challenge both to the scientific investigator and to the practising clinician, for its aetiology is incompletely understood and its almost invariable accompaniment—mental retardation—constitutes a family tragedy.

The onset is in the first two years of life, usually at about six months. In about half the cases (the cryptogenic group) it is the first untoward event in the infant's life. An apparently normal infant starts to have attacks of sudden flexion involving the neck, trunk and thighs; the arms are pushed forwards ('as if punching') or laterally with elbows flexed, and the eyes may turn upwards. Sometimes the neck is extended instead of flexed. A cry may accompany the spasm or immediately follow it. In speed, the spasm may be as rapid as a true myoclonic jerk or somewhat slower. After a few seconds' general relaxation the spasm is repeated, and a bout of spasms usually consists of between 4 and 20. Several bouts occur daily. It is not surprising that the condition is often not recognised as epileptic for several weeks and is diagnosed as abdominal colic, intussusception or Moro reflex, or even as attempts to sit up. However, the coincident rapid mental regression eventually makes it clear that some cerebral insult has occurred. The child loses contact with his environment, sometimes to such an extent that he is thought to be blind, and loses his social responses, though his locomotor functions may be fairly well preserved.[6]

The remaining cases—the symptomatic group—usually have a fairly definite aetiology. Most of these children have suffered prenatal or perinatal brain damage; their histories contain the events often associated with cerebral palsy (extreme prematurity, antepartum haemorrhage, difficult delivery and neonatal apnoea, cyanosis and feeding difficulty). Others are examples of post-encephalitic brain damage, tuberose sclerosis, phenylketonuria and cerebral agenesis with absence of the corpus callosum. Microcephaly is common. Hemiplegia and spastic quadriplegia occur, although generalised hypotonia and hyporeflexia are the commonest findings both in the symptomatic and cryptogenic groups. And, in contrast to the cryptogenic children, in this group mental retardation dates from birth.

Once the condition is suspected, EEG examination is required, for this is one of the few conditions where an EEG is almost diagnostic. It is nearly always grossly epileptic, and, since the alternative clinical diagnoses are non-epileptic, this finding is sufficient confirmation. However, in about half the cases the record also has the characteristics of hypsarrhythmia[5]—namely, chaos and abnormally high amplitude—

and this is rare in other types of epilepsy at this age.[1] It should be noted that hypsarrhythmia is a term which describes an electroencephalographic appearance and is not synonymous with the clinical term, infantile spasms.

Treatment has until recently been unsatisfactory, for the usual anticonvulsants are ineffective. However, in 1958, Sorel and Dusaucy-Bauloye[9] reported dramatic improvement in the epilepsy, the EEG appearances, and the mental level after treatment with ACTH. The epileptic and EEG improvement during treatment have been confirmed by others [2 3 8], but there is not yet sufficient evidence to judge whether ACTH has a more permanent effect. There is a natural tendency for spasms to disappear and the EEG to improve[4 7], and the results of hormone treatment must be judged against this background. The reported improvement in mentality is also difficult to assess. A small minority of untreated children reach a normal mental level after cessation of spasms. It is therefore necessary to demonstrate mental improvement in a substantial proportion of treated cases before the improvement can be ascribed to treatment. Published reports so far have given information which is inadequate for assessment of the frequency and extent of mental improvement. Apparent temporary improvement may merely represent steroid euphoria.

Sorel's discovery is leading to a fascinating new field of investigation and speculation and a more hopeful method of treatment. As a first step early and accurate diagnosis in the individual patient is important.

B. D. BOWER

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Akinetic Mutism

OBSERVATION of a patient with a cystic cranio-pharyngioma led Cairns and his associates in 1941 to describe the condition which they named akinetic mutism. The name implies lack of speech and movement, but the clinical state goes further than this.

The patient sleeps more than normally, but is easily roused. Though he makes no sound and is mute, his eyes regard the observer steadily or follow the observer's movements. His attention may be diverted by sounds. If he is not completely mute he speaks only in whispered monosyllables. There is no restlessness or negativism. A painful stimulus produces reflex withdrawal or an attempt to remove the stimulus, but it is without emotion. Although the patient swallows readily he has to be fed, and he cannot appreciate the appearance, taste and texture of food. He is doubly incontinent. The onset is usually gradual and the condition fluctuates in intensity. When incomplete, it may be interspersed with periods in which there is limited speech and voluntary movement. It is important to emphasise that neither drowsiness nor paralysis are a feature of the syndrome. If the subject subsequently recovers, he has no memory whatever of the episode.

The state is relatively uncommon. Besides its occasional appearance in cases of third-ventricle lesions, it has been seen in patients with pineal and brain-stem tumours, following ventriculography with severe obstructive hydrocephalus, after head injury, and in other conditions. Cravioto and others (1960) have reported no less than 7 patients with basilar artery thrombosis in whom akinetic mutism was present. It is perhaps surprising how relatively rarely the syndrome is seen, considering the frequency of lesions in the upper and lower brain stem. Analysis of the areas of damage in the cases described by Cravioto *et al.*, compared with another 15 patients suffering from basilar artery thrombosis without akinetic mutism, showed that the major distinction between them lay in the greater degree of tegmental destruction in the akinetic cases.

Cairns and others (1941) concluded that the symptoms of akinetic mutism were probably due to interruption of afferent impulses to the cortex, not only of those related to exteroceptive and somatic proprioceptive sensibility but also those of visceral sensibility. In a later paper, Cairns (1952) wrote that there must be interference with 'some hypothalamic-thalamic mechanism which can quickly activate or suppress a large part of the cortex'.

Diffuse aggregations of nuclei may be found among the fibres and cells of the dorsal brain stem from the medulla to the diencephalon. They are known as the brain stem and diffuse thalamic reticular formations. These nuclei receive exteroceptive, proprioceptive and visceral sensory impulses, and exert an activating influence on the cortex. The latter aspect of their function, combined with that of the central grey matter, the subthalamic, hypothalamic and certain thalamic nuclei, constitutes the reticular

activating system. This system affects the state of consciousness and the individual's response to his environment. It is a key point in the integration of the nervous system, but this does not mean that it displaces the cerebral cortex as the highest level of integration in the Jacksonian sense. Cortical activity is very closely dependent on reticular activity, an activity over which it can, nevertheless, exert control. No one area of the brain can therefore be regarded as the seat of consciousness.

The preparation named 'encéphale isolé' by Bremer (1935, 1936), in which the brain stem of a cat is sectioned below the medulla, exhibits a sleeping and waking rhythm, and responds to its environment without emotion. The resemblance to akinetic mutism is striking, although the electroencephalographic tracings are not identical. If, by means of a stereotactic apparatus, a lesion is now placed in the reticular formation of such a preparation, particularly in the periaqueductal area, the state of the animal becomes that of the 'cerveau isolé' of Bremer (q.v.) in which the cut is between the superior and inferior colliculi, and in which the animal appears to be asleep and no longer responds to its environment (Lindsley *et al.* 1949). In 1936, Ingram, Barris and Ranson produced a syndrome in cats which they named catalepsy, in which lesions in the caudal hypothalamus and upper mesencephalic tegmentum caused persistent loss of activity and suppression of emotion, among other things. Bailey and Davis (1942) and Adametz and O'Leary (1959) compared the state produced by lesions of the periaqueductal grey matter in cats with human akinetic mutism. The validity of such comparisons is questionable, but such experiments at least support the conclusion that the essential lesion lies in the reticular activating system.

In their review of akinetic mutism in 1958, Daly and Love point out that the clinical evidence also favours a lesion in the central mesencephalic tegmentum. They emphasise the integrity of the lemniscal system and corticospinal motor tracts. They too conclude that the essential defect is in the reticular activating system, but we have yet to discern more precisely in what part. The system is not a unified whole (Brodal 1957), and we are but on the threshold of discovering its anatomico-physiological relations. Bond and colleagues (1957) concluded that akinetic mutism was not the consequence of damage to specific nuclear masses, but that the lesions were 'so situated as to interrupt major rhinencephalic-hypothalamic and mesencephalic reticulo-thalamo-cortical projections'. This is in agreement with Cairns' conclusions, but the last word has not yet been said.

PETER H. SCHURR

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Second International Study Group on Child Neurology and Cerebral Palsy

Oxford, 1960

THIS year's meeting, from 11th to 17th September, was again held at St. Edmund Hall, Oxford. There were more people than in 1958—eighty-five registered this time, compared with sixty-five—but more of the participants were part-time, and the total group rarely exceeded sixty at any one time.

About three-quarters of the 1958 members came again, and, of those who did not, a number were prevented by illness. Among the new members were Harry and Ruth Bakwin and Richmond Paine from the U.S.A., Edmund Frühmann from Vienna, Norah Gibbs, O. Zangwill, Jack Tizard, Tony Barnett, and the Bobaths from the U.K., to mention but a few.

The programme included the physiology of the spinal cord, to prepare us for a visit to Stoke Mandeville Spinal Rehabilitation Centre; a session on neurochemistry arranged by John Cumings; a start on tackling the problem of assessment of results of treatment, including the use of polymyographic studies; a day on the parietal lobe and disorders of sensation (other than hearing) in the cerebral palsied; a discussion on 'What parents want from doctors'; and a review of the Little Club report on the neurological testing of the newborn infant, which had been drawn up at a preliminary meeting in Groningen in July.

The papers were in general better than in 1958, there was more sessional time for discussion, although some speakers thoughtlessly overran their allotted time, and there was much discussion apart from the sessions. It has been suggested that there should have been more about the causation, pathology, treatment, psychology and emotional aspects of cerebral palsy. Many letters of thanks to the N.S.S. have been received and one member sent a donation. It is clear that this meeting was found to be informative and enjoyable. Some remarked especially on the fresh impetus they have received from it.

The 1960 meeting revealed a remarkable advance in the maturity of the discussions. To take one example—the subject of terminology and differentiation of types of cerebral palsy—two years ago the discussion after the clinical demonstration evoked almost vituperative disagreement; in 1960 there was much less discussion, because there was much more agreement on the use of words and the labels to attach to the various cases. In the discussion on terminology, the area of disagreement proved to be fairly limited and it was not difficult to come to an agreement that each of the various classifications was proving useful in clinical practice and therefore we could each go on thinking in the hope of exchanging further news some time in the future. This great increase in capacity to discuss without confusion, and in agreement on clinical labelling of cases

themselves, alone would make the meeting well worth while.

When the next Study Group meets at the same place in two years time, we shall hope to hear more from the educational field.

Publication of papers read at this year's Study Group, and the discussions which followed them, will commence in *Bulletin* Vol. 2, No. 4 and will be continued in subsequent issues.

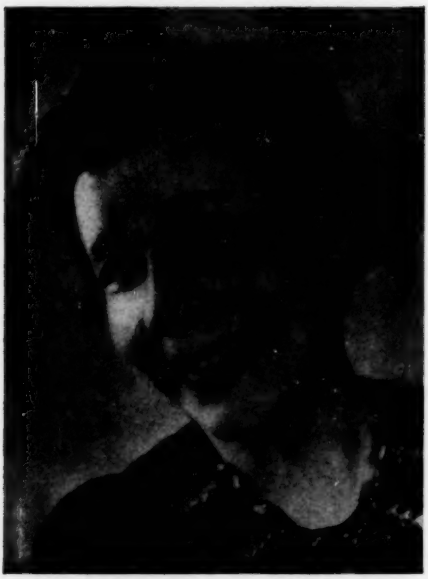
R. C. MAC KEITH

NOTE: *Nobody who attended this Study Group would like the occasion to pass without some mention of the astonishing skill, energy and tact of its organiser, Ronald Mac Keith.—E. Clayton-Jones.*

OBITUARIES

Mme. Stella I. Albitreccia

THE sudden and unexpected death of Mme. Albitreccia in Paris on June 10, at the age of 59, came as a great shock to everyone who knew her. It is a great personal loss as well as a professional one. She was known to many people in this country through the brilliant pioneering work that she did on spatial perception and disorders of the body image in brain-injured children. Her work in this field led her to devise teaching methods and a programme of special education designed to train children with disorders in these areas. Those of us who worked with her will miss her tremendous vitality, her passionate devotion to her work, and the affection and understanding with which she treated handicapped children and their families.



Mme. Albitreccia in 1955

It is amazing to realise that Mme. Albitreccia's pioneer work with cerebral palsy did not begin until she was well over 40. She came from a medical environment, but in her early years her diverse talents and interests led her from teaching into journalism. She was born in Bihac, Bosnia (then part of the Austro-Hungarian Empire), where her father was the director of the City Hospital and the City Health Commissioner. She was active in the Red Cross during the First World War. For two years she worked in a mobile soup kitchen and often fed as many as 700 destitute women and children a day. Mme. Albitreccia's brother, Dr. Victor Koerbel of New York, tells me that at the same time she fed all the dogs in the area who had become savage through neglect. These wild animals became quite tame in her presence, and would wait quietly to be fed. They apparently felt that they were in the presence of someone who understood and was trying to help them. Something of this same spirit was apparent in her work with children, who instinctively seemed to sense that she was a friend who would understand their difficulties and try to help them to overcome them. Throughout her life her many friends came to feel this great wisdom and understanding and deep sympathy she felt for people as well as animals.

At the end of the war her family moved to Vienna. There Mme. Albitreccia completed her language studies, and passed the State Board examinations for teachers of English and German. She taught English

until 1933 at the Urania, a sort of 'popular' university. At the same time she worked as a freelance journalist for several Viennese and foreign newspapers. She was concerned with problems of maternal and child welfare, and wrote historical and sociological essays. She was always interested in art, and became an art critic for several newspapers. She often came to England at this time, and at one time joined the Salvation Army as an observer. This resulted in a series of articles about its work in a Viennese newspaper.

In 1933 she became interested in logopedics and phonetics. She studied under Dr. Hugo Stern, who was in charge of the Logopedics and Phonetics Department of the Second University Clinic for Diseases of the Ear, Nose, and Throat, of the University of Vienna. She became Dr. Stern's assistant, and became deeply interested in aphasia. Studying this subject led her into research on the various methods and approaches used in treatment.

In 1938 she married Prof. Antoine Albitreccia. By mere chance she happened to be in Paris looking for a flat, and thus missed the *anschluss* by only a few days. She barely escaped the Nazis who had come to look for her because she had been an art critic for a Danzig newspaper which was anti-Nazi. She remained in Paris, and worked under Dr. Ramadier at the Hôpital de la Pitié, doing speech and voice correction in the E.N.T. department. She also became a teacher of English and German at the Collège Sevigné, and later a school psychologist. She continued to teach normal children until just a year ago, for she felt that this was important in her work with handicapped children. It helped her to gain perspective on the questions posed by the abnormal, and to understand how the normal child learns.

At the fall of France she and her husband were active in the de Gaullist underground. They hid many people and arranged for their escapes to the coast and then to England. She was arrested in 1942 by the Gestapo; presumably someone she had helped to escape had informed them of her activities. She was beaten and tortured, and was thrown on a pile of corpses in a courtyard as dead. Her astonishing vitality and courage stood her in good stead now. She came to under a sailcloth cover on top of the pile of bodies, and through another extraordinary chance was able to escape from Gestapo headquarters with the charwoman who was just leaving. She found refuge with a friend, who hid her and later helped her to escape into the unoccupied zone. For the rest of the war she and her husband lived in Albi, still active members of the Resistance. She was a stretcher-bearer and radio-operator. She and her friends picked up parachutists and parachuted supplies, hid refugees, blew up trains and depots, and harassed the enemy in every way. To many of the children that she hid she became a second mother, and they were devoted to her for the rest of her life.

When Paris was liberated, her husband was transferred back there to join de Gaulle's general staff. Her husband was to have been sent to Austria in a high governmental post, but he was tragically killed in a motor accident in August 1945. Mme. Albitreccia was severely injured in the same accident, sustaining 23 fractures as well as a brain injury which caused her to be aphasic herself for a time. She felt that being brain-injured herself gave her a deeper insight into the problems of the aphasic patient and also helped her to know which approaches would work best in treatment. For a time it was feared that she might never walk again. Once again, though, her incredible courage and determination carried her through. That she managed to overcome all the pain and hardship and go on to do the fine work she did remains a near-miracle to those who knew her.

In 1945 she joined the Aide aux Enfants Paralysés in Paris. There she worked with Dr. Auguste Tournay, who had begun this first centre for the rehabilitation of children with central nervous disorders. Mme. Albitreccia undertook the re-education of sensory disturbances and speech disorders.

She worked closely with Mlle. Marguerite Mellerio, who is now the director of this centre and in charge of motor re-education. Madame Albitreccia was herself particularly concerned with the fields of intelligence testing and pre-scholastic training.

During the last few years she was occupied with perfecting the techniques which were used in her special class at the Aide aux Enfants Paralysés. Her class aroused much interest in teaching and medical circles and was visited by many people from all over the world.

She put together a battery of tests which were designed to diagnose disorders of the body image and spatial perception in children with cerebral lesions. Her theory was that the child who could not move properly would fail to acquire a correct mental picture of his own body, and that this lack would prevent him from acquiring the correct spatial orientation so essential for learning. In her class, through the use of tests and specially designed exercises, it became obvious that there is a correlation between disorders of the body image and spatial orientation and the many physical and scholastic problems present when there is a need for precise organisation and localisation. This becomes apparent in speech, writing, motor activities, phonation, and spoken and written language. Mme. Albitreccia divided the children into groups of four to five, and gave them special exercises according to individual needs and rates of progress, all of which were aimed at providing sound scholastic preparation. An interesting result of this training in body image and spatial perception was the drawings that Mme. Albitreccia got the children to do. As an example, a boy with hemiplegia drew a man with only one side; ignoring the paralysed limbs. After six months a notable progress could be seen in the child's drawings as he became conscious of the other side of his body. Scholastically the child progressed as he became aware of both sides of his body.

Madame Albitreccia has left us the legacy of her work, which is a serious and profound contribution to our understanding and treatment of cerebral palsy. Those of us who were trained by her and worked with her have many memories of her tremendous energy and her brilliant searching mind, combined with great charm, wit and gaiety. Being near death so many times seemed to give her a tremendous vitality and zest for living. She was always interested in many things, and art remained one of her main interests throughout her life. Her wide circle of friends loved her for her wit and charm and for the interest and sympathy she invariably showed. It is difficult to think of her with sadness, for she was always gay and optimistic and always maintained the sense of humour that had taken her through so many dark days. All of us who knew her, whether professionally or personally, owe her a great debt.

JUDITH M. HILLELSON

Melanie Klein

Melanie Klein was born in Vienna, of Jewish parents, in 1882, and she died on September 22, 1960. Early marriage and the care of three young children prevented the realisation of her ambition to become a physician, but she became intensely interested in psychoanalysis and was able to pursue its study under Ferenezi and Abraham in Berlin and Budapest. After working for some years in Berlin, she came to London in 1926 at the invitation of Ernest Jones and remained there for the rest of her life. Long before her death, at the age of 78, she had come to be recognised as one of the most important of those who had extended the original boundaries of Freudian theory and practice, and in her own field of child analysis her reputation was supreme. Her chief books are *The Psycho-Analysis of Children* (1932), *Contributions to Psycho-Analysis* (1948), *Developments in Psycho-Analysis* (1952), *Envy and Gratitude* (1957), and *The Narrative of a Child Analysis*, which was in course of publication at the time of her death. The essay reviewed below forms a convenient introduction to her work and should stimulate many to read more of it.

MELANIE KLEIN began her psycho-analytic work with children at the end of the first world war as an assistant to Ferenczi in Budapest. Her lectures in London in 1925 and 1927 achieved a notable success, and at about this time she settled down there, to become an influential member of the British Psycho-analytic Society. The publication in 1932 of her book, *The Psycho-analysis of Children*, marked an important step forward. Freud had undertaken the analysis of a five-year-old child, 'little Hans', and Hug-Hellmuth, Anna Freud and others had made use of play methods in the analysis of young children, but Melanie Klein's play techniques were more advanced than those of her predecessors in this field and have exerted a considerable influence on contemporary methods.

Her theories quickly gained some support, perhaps especially from Ernest Jones. Freud, on the other hand, was sceptical of them, and they have remained controversial. She has argued that the most critical experiences occur in the first six months of life, and she has interpreted reactions to frustration, such as rages, withdrawal of interest and mood changes, in terms of the mechanisms revealed in the psycho-analysis of adult patients. Frustration, discomfort and pain, she has supposed, are experienced in the first few months of life as persecution. This, 'the paranoid position', in which destructive impulses and persecutory anxiety predominate, is the first of two main phases in the first six months of life. In the second, 'the depressive position,' introjection occurs, and the infant suffers anxiety lest the introjected object be harmed or destroyed. The super-ego has its origins in the fifth or sixth month of life and Oedipus conflicts can also be traced back to this time. The infant may refuse the breast, she has suggested, because he has not been able to establish trust and love towards the mother.

Her work has been criticised on the grounds that she has attributed to infants much more developed capacities for projection, introjection and transference than her observations warrant. Current researches into the mental activities of infants now suggest, however, that there is much more in her theories than has yet been generally admitted, although the terms in which they have been expressed may require revision if they are to be incorporated into an ecletic psychopathology. Her emphasis on the first few months of life has a very modern ring.

In *Our Adult World and its Roots in Infancy*,* an essay of 6,000–7,000 words, she has given a brief but comprehensive statement of her findings and theories in terms comprehensible to readers who are not psycho-analysts.

D. RUSSELL DAVIS

**London:* Tavistock Publications, 1960, 15pp., *3s. 6d.*

BOOK REVIEWS

Handbook of Speech Pathology*

Edited by LEE EDWARD TRAVIS, PH.D.

Reviewed by T. T. S. INGRAM, M.B., M.R.C.P.E., D.C.H.
Lecturer in the Department of Child Life and Health, University of Edinburgh.

This massive volume contains chapters written by twenty-seven different American authors on their special interests in 'Speech Pathology'. The distinguished editor has tried to cover the whole subject of speech and language disorders in childhood and adult life. Aetiology, pathology, clinical findings, psychological complications, treatment and prognosis are all considered.

The book is divided into four parts. The first is devoted to *Basic Considerations in Speech Pathology*, the second to *Speech and Voice Disorders Associated with Organic Abnormalities*, the third to *Speech and Voice Disorders Unrelated to Organic Abnormalities*, and the fourth and last to *Psychotherapy and Speech Therapy*. In a short review it is impossible to criticise all the 33 chapters in any detail.

Inevitably there are great variations in the value of the individual chapters as expositions of their subjects and in their literary merit. Perhaps some authors were too overcome by the first few lines of the first chapter, on *The Development of Speech*, to do themselves justice, while others rose to the occasion. 'In the modern world some two-and-a-half billion people pour out words in astronomical numbers. Daily they talk to each other in person, and in certain areas of the world over the telephone, on radio and on television. Uncounted trillions of additional words are used in the covert processes of remembering, thinking and imagining.' Yes, this is a very American book.

It was a good idea to give up the second chapter to *Terminology and Nomenclature*, but not all the medical terms in the glossary are precisely defined. The chapters on *The Acoustics of Speech* and *Instruments of Diagnosis and Research* appear disproportionately long, but are clearly written accounts which will be useful for reference. The chapter on the *Neurophysiology of Speech* cannot be considered seriously as an adequate account of the complex neurophysiological changes which underlie speech production. It consists largely of vague generalisations about behaviour.

In the second and third parts some of the chapters describing speech disorders are excellent and some are disappointing. Those on *The Pathology and Treatment of Deafness*, by Goodhill, and on cleft palate, by Koepp-Baker, are well written, well illustrated and deal with their subjects systematically and comprehensively. The chapters on functional disorders of articulation, by Margaret Hall Powers are also informative, well designed and readable, though after reading them one's doubts about the 'functional' nature of some of the disorders of articulation that she describes are increased. The review of *Speech Problems of the Mentally Retarded* gives an adequate account of the prevalence and prospects of treatment of speech disorders in the mentally handicapped, but says too little about their aetiological and clinical characteristics. Perkins and Garwood do not seem to come to grips with the problems of speech therapy in cerebral palsy in their 15-page chapter, though they append an acceptable bibliography to it.

There are three chapters on aphasia in

* *London:* Peter Owen, 1959, pp. 1096, 84*s*.

adults by Eisenson and two on aphasia in children by Myklebust. Even allowing for the difficulties of writing about aphasia in short chapters these authors' names promise authoritative and readable accounts of their subjects. Unfortunately all five chapters are inadequate and will probably have to be replaced in subsequent editions. Neither author has succeeded in presenting a comprehensive, systematically written chapter. For example, in *Classification and Examination Procedures* of aphasia in adults the various methods and difficulties of classification are not fully discussed, and the methods of examination are not sufficiently described for the reader to see how enough clinical findings were elicited for classification to be possible. The section on *Motor Capacities* in the chapter dealing with the diagnosis and training of children with aphasia, by Myklebust, also leaves much to the imagination. 'The aphasic child is delayed both in the age of sitting and walking as compared to the normal (Myklebust 1954)' is but one of many statements which require elaboration or deletion.

Stuttering, or stammering, receives due attention, for there are four chapters on it by well-known authors. Van Riper writes about *Symptomatic Therapy for Stuttering*, Johnson on *Perceptual and Evaluational Factors in Stuttering*, Travis on *The Unspeakable Feelings of People with Special Reference to Stuttering*, and Ainsworth on *Method for Integrating Theories of Stuttering*. None of these chapters presents a systematic account of the aetiology, pathology, manifestations, course, treatment and prognosis of stammering, but each one does present the point of view of an experienced 'speech pathologist'. All the articles are well worth reading and it is perfectly possible to agree with the one which most nearly represents one's own views without disagreeing with Ainsworth too drastically.

The last part of the book is devoted to three chapters on *Psychotherapy and Speech Therapy*. The first is *The Psychotherapeutic Process*, by Travis, the second *Play Therapy, Psychodrama and Parent Counselling*, by Wolpe, and the third *Group Structure in Speech Therapy*, by Backus. The first shows a very strong psychoanalytical orientation, and both describe case-histories in considerable detail.

The book is valuable for a number of reasons. Firstly, it is by far the most comprehensive reference book dealing with speech defects in adults which has yet appeared. Secondly, it does present a picture of the work of American speech pathologists at the present time. Thirdly, it should make people interested in speech disorders in Britain a little more concerned than they are about the future of speech therapy. Lack of medical interest in the United States has led many speech pathologists to attempt to diagnose and treat their patients without adequate medical aid. The dangers of this are well illustrated in some of the chapters in this book, which would have been immensely improved if the co-operation of trained medical men had been available to describe pathological, aetiological and clinical findings in broader perspective. Yet already in Britain speech therapists are carrying much of the burden of the diagnosis as well as the treatment of their cases, because of medical neglect of speech disorders.

The Chesty Child

London: Published by the Chest and Heart Association, 1960, pp. 96, 8*s.* 6*d.*

The chesty child remains one of the common problems of paediatrics, not only for the parents and family doctor but also for the school, the hospital, and indeed the community.

A symposium on this subject was held in London in June 1959 by the Chest and Heart Association, and the proceedings have now been published in this booklet, whose 96 pages cover many aspects of the

personal and community care of such children. An index would have increased its value.

The 16 contributors include a parent, a teacher, general practitioners, school doctors, various hospital specialists and social workers, and each approaches the problem from a different angle. It is therefore interesting that all stress 4 or 5 common factors of importance in general management, and emphasise that consideration of the whole child and his environment is of paramount importance.

Stress is rightly laid on the need for accurate diagnosis of the cause of the symptoms in each child; coughs of nervous origin and those arising from conditions outside the chest itself need different management from those arising in the lower respiratory tract. Tuberculosis as a cause of chestiness in childhood is now a thing of the past in this country, but parents do not know this, and it is important to dispel their anxiety about this disease, together with other outmoded fears of draughts, chills and similar supposed evils. Repeated explanations of the cause and effect of symptoms play a major part in the management of every patient. This is time-consuming but helps parents and child alike to shed some anxiety, as well as to co-operate more intelligently in treatment.

Infective factors are the most amenable to prevention and treatment, and children with severe bronchiectasis are a minority these days, although chronic bronchitis is still a problem. Fibrocystic disease, now relatively common, receives insufficient mention as a separate entity. It is surprising, too, to find no mention of long-term antibiotic therapy, but the emphasis of the symposium is rightly on the wider aspects of allergy, psychological, educational and social factors. The value of physiotherapy is stressed, and so is the need to provide this in schools to prevent waste of educational time in attending clinics.

Allergic factors are known to be difficult to assess but are often important in the older child, especially when there is a strong family or personal history of allergy. Careful study of the environment is noted as being more rewarding in eliminating allergens than skin testing. Moulds and spores are significant allergens in some children who show marked climatic variation in symptoms, and more research is needed in this field.

All the contributors lay stress on the complex psychological factors involved, which we do not yet pretend to understand fully. The interplay of emotional factors between patient, parents and siblings varies from family to family, and vicious circles of unconscious anxieties, dependence, aggression and guilt are frequently set up. The child's regression to immature dependent relationships, as an escape from a world too challenging for him, is often fostered by parental wishes to prolong the stages of dependence. Anxiety over physical symptoms, especially dyspnoea, in both child and parents, may stem from earlier unconscious fears, and parental disharmony may be manifested through the child and his symptoms.

An attempt to understand the individual psychological mechanisms at work is essential before treating each patient, and this calls for co-operation between family doctor, hospital specialists, school doctors and the teachers concerned, as well as the parents and the child himself. This, too, involves considerable time and trouble but is most worth while.

The chapter on exercise, sport and games emphasises the benefits to personality development arising from the acquisition of self-control. Mastery in some physical skill involving control of his body gives the child a sense of enjoyment and confidence which is of value beyond the skill itself. Sport also gives an opportunity for exercise, release and control of aggressive instincts, and for these and other reasons should not be denied to the child with respiratory difficulties. A sense of inferiority already exists in such children, and limiting their activity increases it unnecessarily. There is little danger in allowing these patients to push themselves to the limit of

their own capacity. Others may, at first, need stimulating to further physical effort to release them from the fears engendered by an over-watchful home environment.

Hobbies and games at which they can excel are important pursuits to be encouraged to help overcome the frustrations of limited physical activity in the more handicapped children. Integration of these enterprises through the school is of great value to the child.

Much more co-operation is needed between the school and the various medical and social agencies concerned, to prevent the enormous amount of absenteeism and loss of education from which these children suffer. The benefit of the various types of special education are discussed, but not enough stress is laid on the value of residential schools, especially in asthmatic children. Much research is needed on this aspect of the problem alone, for all concerned are familiar with the remarkable benefits of separation from home, whether to hospital or convalescent home, to relatives, or to a boarding-school for asthmatics. The valuable experience with English asthmatic children in Switzerland is described, but equally good results are obtained in boarding-schools in this country. How far allergic, emotional or other factors contribute to the almost universal improvement is unknown. So, too, are the physical and psychological differences which presumably exist in those very few children who fail to derive much benefit from such separation.

Brief mention is also made in this booklet of cardiac conditions, congenital or rheumatic, as they affect the child in his environment. The need to avoid invalidism is stressed, and also the benefit of more detailed reports from specialists to family doctors, in the rapidly developing field of cardiac surgery.

Little mention is made of the importance of better education of doctors in all the factors involved, especially in an understanding of the interpersonal relationships in family life. Clearly this would result in more effective prevention and treatment of chesty children in the earliest stages of their disability. Wider education of the general public in such matters might also be beneficial.

Finally, as has been seen, research is urgently needed in many aspects of the origins and treatment of the child with chronic respiratory disease, which now, with behaviour problems, forms the major part of both home and hospital paediatric practice. The school doctor is favourably placed to investigate many facets of this important condition.

CHRISTINE COOPER

Paediatric Neurosurgery

Edited by IRA J. JACKSON and RAYMOND K. THOMPSON.

Oxford: Blackwell Scientific Publications. *Springfield, Illinois:* Charles C. Thomas, 1959, pp. 564, £6 5*s*.

The publishers' intention here was to provide a textbook in which the opinions of its twenty-four contributors would represent a cross-section of current thought on paediatric neurosurgery. However, unless considerable licence is given to the editors, such a large number of authors tends to produce a lack of balance and cohesion. Different aspects of the subject do not form watertight compartments, and it is necessary to have a thread of continuity, particularly in the matter of treatment. In this book one feels that some specialists have over-emphasised their contribution while a few important aspects of the subject slip by almost unnoticed or fall between the chapters.

For the paediatrician and general practitioner this book is a display of neurosurgical wares. It describes new advances in general terms and is of value in guiding the physician or surgeon who may be

responsible for the care of a patient before and after definitive neurosurgery. One must remember, however, that fundamentally the subject is Neurology, and there are many conditions in which the main line of therapy is medical, and to which the surgical contribution is small, however important it may be. Such conditions are better considered from a broader point of view than that of the surgical aspect alone. Nevertheless, it provides a source of references to surgical literature which may not be regularly perused by medical readers. For the experienced neurosurgeon the book has very little to offer. He seeks information on points of rarity, or asks for the experience of those who see more of a particular problem than he does. The book is written in terms which are too general, and at times too elementary, to be of help to him. For the trainee neurosurgeon who is at the beginning of an arduous period of apprenticeship, and for the house-surgeon and registrar who wish to read generally about the subject, this book is a most excellent introduction.

Particularly noteworthy are the chapters on intracranial tumours and on trauma, which cover the field admirably, though a few more illustrations of the pathology of tumours in children would make the work of more value to general pathologists and to physicians interpreting the reports of specialists. There are some surprising omissions—cerebral palsy is hardly touched on, and the indications and uses of hemispherectomy are not discussed at all. The rehabilitation and physical treatment of paralysed children is hardly given a reference, and one is advised to ask a urologist about the care of children with chronically paralysed bladder and sphincters. The reader is interrupted by a large number of errors which should have been corrected in proof. These are not only typographical, but occasionally involve mis-statement of fact.

There are surprisingly few textbooks dealing with the surgical aspects of neurology, and a work devoted to Paediatric Neurosurgery is a welcome addition to the literature. Coming within a few years of the production of another book on the same subject by the same publisher, comparison is inevitable, and to some extent the two books are complementary. Ingraham and Matson's *Neurosurgery of Infancy and Childhood* is a museum of the material of one of the largest centres of paediatric neurosurgery. Jackson and Thompson provide a background for the detail of the earlier book. This is a volume which should be on the shelves of most paediatric departments but its appeal may be limited.

PETER H. SCHURR

Neurology of Infancy

By ANATOLE DEKABAN. *London:* Bailliere, Tindall and Cox, 1959, pp. 388, £4 16*s*.

A comprehensive textbook dealing specifically with the neurology of infancy has long been needed. Dr. Dekaban has not quite provided it. He has written a quite remarkable and, at the same time, a rather frustrating book. His aims were 'to provide paediatricians, neurologists, child psychiatrists and physicians participating in well-baby care with anatomical and developmental landmarks of the maturation of the central nervous system during the first two years of life' and to 'outline the principles of pathology, pathogenesis and clinical manifestations of neurological disorders occurring during infancy'.

He attempts to reach his first aim in the first two chapters, in which he reviews the milestones of anatomical and neurophysiological development in the first two years, and correlates clinical and neuroanatomical findings. The illustrations of neuroanatomical specimens at different ages are excellent. One's only criticism of this part of the book is that it is too brief and too closely written. Paediatricians,

familiar with the concept of clinical findings changing in children as they mature, will appreciate the closely packed facts which the chapters contain. It is unlikely that the majority of strictly trained younger neurologists and psychiatrists in this country will find them readable enough to be easily acceptable. Their more open minded elders will probably enjoy their reading, though the chapters might, indeed, have been made more attractive by including illustrations of the stages of neurophysiological development to match those of the neuroanatomical findings.

In the remaining chapters the various disorders of the nervous system which affect children under the age of two years are described in a concise and dogmatic fashion. There are chapters on birth injuries, vascular disorders, encephalitis and encephalomyelitis, meningitis, dysraphic states, 'various conditions underlying cerebral palsy', hydrocephalus, epilepsy, mental deficiency, 'exogenous and endogenous intoxications affecting the nervous system', neuro cutaneous dysgeneses, neoplasms, metabolic and endocrine disorders, progressive degenerations of the central nervous system, and primary diseases of the muscular system.

Thus a comprehensive range of neurological disorders of infancy are described. Many of them require and have received fuller consideration individually in larger books than this. It is inevitable, therefore, that the descriptions of the various diseases that are considered should be brief and that the opinions should be dogmatic. Only 17 pages could be devoted to epilepsy and 3 to the pathological, clinical and therapeutic aspects of neurosyphilis. The aetiological factors concerned in birth injury receive relatively scant attention, and the descriptions of the clinical findings in some of the less common types of cerebral palsy are frankly inadequate. The pathology, symptomatology, treatment and prognosis of glioma of the optic nerve can hardly be expected to be fully considered in 11 lines, nor can the neoplasms of the basal ganglia and thalamus be dealt with properly in 4.

But considering the scope of his subject and his ambitious attitude towards it, the author's achievement in this small book is quite remarkable. Most of the important neurological conditions of the first two years of life are adequately described and some of them, particularly metabolic and endocrine disorders, receive quite masterly summaries. The author consistently gives informative references to the various disorders he is discussing even when his descriptions are at their briefest. The American bias of the references he quotes is understandable. It is possible to quarrel with few of his dogmatic statements.

The volume is beautifully printed and produced. The magnificent illustrations, which consist largely of photographs of patients and pathological specimens, show exactly what the author says they do. (This is sufficiently unusual to be remarkable.) The index is adequate.

In short, this book provides a valuable and well set out synopsis of what is known about diseases affecting the nervous system of children under the age of two years, although it is not a comprehensive reference book for neurological diseases of infants in the same way as 'Ford' is a reference book for diseases of the nervous system in older children.

T. T. S. INGRAM

Aids to Neurology

By E. A. BLAKE PRITCHARD.

London: Baillière, Tindall & Cox, 2nd edition, Students Aid Series, 1960, pp. 480, 15*s*.

This is a textbook of neurology in small print on small pages. As such it makes instructive but by no means light reading. Neurology is a relatively precise branch of medicine and the reader of this book will find in it all he needs to enable him to

elicit the history and physical signs and to carry out the special investigations necessary to reach a correct diagnosis in a neurological case. The author believes (p. 333), however, that 'the treatment of all functional disturbances should be in the hands of the psychiatrist in all cases in which it can be reliably inferred that they are not based primarily upon some remediable organic dementia.' What a pity to find so eminent a physician as Dr. Blake Pritchard accepting without protest the fragmentation of medical practice into rigidly segregated specialities! The Cerebral Palsies are adequately dealt with in four pages, but again the implication is that the cases should be dealt with by specialists, so that no real guidance is given about treatment.

This book does well what it sets out to do and can be recommended as a sound presentation of present-day neurology of adults.

JOHN BURKINSHAW

Brain Memory Learning

By W. RITCHIE RUSSELL, C.B.E. *Oxford:* Clarendon Press, 1959, pp. 140, 18*s*.

During his many years of clinical work with brain-damaged people, Dr. Ritchie Russell has explored widely in related fields in search of better understanding of complex aspects of human behaviour, and some of his cullings are presented in this book.

Conventionally enough, Dr. Russell begins with the physiology of neurones and the elaborate organisation of the brain, but before long he has introduced such topics as the effects of concussion and contrasts them with those of focal lesions of the brain: hallucinations; epilepsy; phantom limbs; pain and its control. He makes frequent reference to the emotions and even to what psychoanalysts have to say. He emphasises the importance of facilitation and repetition, and stresses the similarity of thinking and remembering to sensorimotor skills—'As with the simplest neuronal systems the tendency to repeat what has happened before pervades all activity'.

The title, 'Brain Memory Learning', may be daunting, covering as it does three of the most characteristic and familiar, but elusive and obscure features of our species. But the subtitle, 'A Neurologist's View', is reassuring, and this is reinforced by a statement in the preface that the presentation is both personal and speculative. It is refreshingly so, for no attempt at exhaustive and exhausting treatment of the subject is made, and it is the view of a very humane neurologist we are invited to see. Dr. Russell's philosophy is summarised in the final paragraph: 'Clearly if we wish to make human life something more worth while than it now seems to be for many people, we must attempt to comprehend the incredible possibilities for development that are present in every child's brain.' And earlier he says: 'Children should not be expected to make up their own minds in a reliable way, and yet it seems desirable to surround them with all good traditions, beliefs, and customs which will slowly enable them to acquire a capacity for decision of a type which is desirable or beneficial to the community.' If he does not disdain thus to wear his heart on his sleeve, he makes it clear that his mind is firmly in a mechanistic place, for he says: 'It will be generally agreed that the earliest reactions of the infant to his mother can be explained as an elaboration of the behaviour of simple neuronal networks.'

This unusual coupling of science and sensibility is further exemplified by such remarks as, 'the amazing complexity of the neuronal system involved in remembering should make us less critical of those who become absent-minded or forgetful of the names of their friends.' And so one can take double pleasure from Dr. Russell's book, reading it as an introduction both to what makes the brain tick and to what makes a particular neurologist tick.

M. L. JOHNSON ABERCROMBIE

Maladies héréditaires du Métabolisme chez l'Enfant

By M. LAMY, P. ROYER and J. FRÉZAL. *Paris:* Masson, 1959, pp. 260, 36 *N.F.*

In the past the inborn errors of metabolism have too often been considered medical curiosities of little practical importance. They have been regarded as suitable diversions for medical meetings and used to dismay examination candidates. Today, however, the vast and ever-growing mass of information concerning the biochemical functions and malfunctions of the human body far exceeds the capacity of the physician to absorb it and he is left breathless by the rapidity with which new knowledge accumulates. Not only have the well-known 'inborn errors' become complicated beyond recognition but almost daily new ones are discovered or old familiar diseases are found to be due to enzyme deficiencies. It is more than likely that in the near future abnormal metabolism will be held accountable for an appreciable number of cases of cerebral palsy. Thus this formerly small group of rare conditions has grown into a major branch of medicine and one which, complex though it may be, no physician can afford to ignore.

Hitherto there has been a shortage of good books in this field and the newer textbooks on metabolism often give insufficient space to paediatric aspects. This book from the Paris medical school is therefore a welcome addition to the slender library on the metabolic disorders of childhood. It is not an exhaustive or comprehensive survey but rather a treatise on selected conditions of which the authors have had special experience or which illustrate particularly well the concept of inborn errors of metabolism. Thus the reader will find 18 pages on congenital suprarenal hyperplasia but none on familial cretinism; there are 7 pages on alcaptonuria, but Hartnup disease and maple syrup disease are not discussed; there is no chapter on disorders of haemoglobin and pigment metabolism. The conditions which are included are dealt with at considerable length and the discussions bear witness to the authors' wide experience. The treatment is by no means parochial, however, and there is frequent reference to work in other countries. Perhaps the authors have been wise in restricting the scope of their book, for so many conditions are now regarded as metabolic disorders that they could not all have been considered in such detail.

The conditions discussed are galactosaemia, the glycogenoses, anomalies of aromatic amino-acid metabolism, congenital suprarenal hyperplasia, anomalies of tubular transport, vitamin-resistant rickets, the de Toni-Debré-Fanconi syndrome and cystinosis, diabetes insipidus, mucoviscidosis, the sphingolipidoses, gargoylism, and spontaneous hypoglycaemia. The arrangement of chapters and subjects seems unsystematic and rather haphazard but perhaps this is merely because it is different from the usual practice. Thus the chapter on anomalies of tubular transport includes cystinuria and renal glycosuria, but vitamin-resistant rickets and nephrogenic diabetes insipidus are considered in separate chapters. Hypophosphatasia and familial vitamin-resistant rickets are included under the same heading, as has been customary hitherto, but surely it is time they were separated since the mechanisms of their production are probably entirely different.

The chapters on the de Toni-Debré-Fanconi syndrome and cystinosis, on fibrocystic disease of the pancreas (mucoviscidosis), and on gargoylism are particularly good. There is a stimulating preface by Prof. Robert Debré, who has himself contributed so much to knowledge of these disorders. There is also a useful introductory chapter on genetics and the mechanisms of metabolic disorders, and one on the heredity of diabetes mellitus. It is pleasing to see the ample list of references at the end of each chapter, but the lack of an index is an irritating feature. The book is paper-bound, as is the French custom, and the paper on which it is printed is not of uniform quality throughout. One can-

not help feeling that a book as good as this is worthy of a better binding.

The greatest value of this book is perhaps the detailed information it gives on the genetics and biochemistry of each disorder. Clinical aspects are adequately dealt with, though the illustrations are too few and of uneven quality. In such a condition as suprarenal hyperplasia, for instance, where photographs can be most informative, there are no illustrations at all. More consideration of differential diagnosis would also have added to the clinical usefulness of the book. For example, the clinician faced with a case of cardiac glycogenosis in an infant will obtain little guidance, for differentiation from other causes of cardiomegaly receives no mention. Management of the various conditions is only briefly dealt with and some important aspects are omitted altogether. Thus in the 10 pages devoted to phenylketonuria there is no mention of the important question of screening newborn infants for this anomaly.

These few criticisms concern omissions which in the reviewer's opinion might have enhanced the book's usefulness to the practising paediatrician. They do not in any way diminish the great importance of this contribution from the Hôpital des Enfants Malades, which will be essential reading for paediatricians, biochemists, geneticists and others interested in metabolic disorders of children, and will undoubtedly remain a valuable source of reference for many years to come.

ROSS G. MITCHELL

New Opportunities for Deaf Children

By I. R. and A. W. G. EWING. *Springfield, Ill.:* Thomas, 1958, pp. 149, 15*s.*

For the past ten years there has been a growing awareness of the high incidence of deafness among the victims of cerebral palsy, of the importance of making an early diagnosis and assessment of the deafness, and of the need to provide special treatment for such doubly handicapped children.

Readers of this *Bulletin* anxious to know something about the problem of diagnosing deafness in infancy and childhood and the way in which deaf children should be managed at home, in clinics and at school could not do better than read this splendid little book.

Published in 1958, a short time before the announcement of Professor Ewing's knighthood and Lady Irene Ewing's death, it is now well known and highly regarded, not only by those for whom it was intended —the parents and teachers of the deaf child—but also by doctors, psychologists, the staffs of maternity units, welfare workers and many others.

Its excellence stems from the authors' profound knowledge of the problems of the deaf child and the deep and practical sympathy for him which permeates every page. Almost everything that is written relates to their own experience and observations, and many of the techniques described with such clarity are of their own devising.

A long section is devoted to case histories, some of them written by the parents and patients themselves. These form a series of impressive illustrations of the way in which this great challenge has been and is being met and mastered through the devotion and enthusiasm of teachers of the deaf, physiotherapists and speech therapists, and particularly by the Ewings of Manchester.

L. F. W. SALMON

Family Clinic

Articles by the *Sunday Times* Medical Panel, edited by DR. BARBARA EVANS. *London: Sunday Times,* 1960, pp. 196, 10*s.* 6*d.*

For the last three years the *Sunday Times* has published a weekly feature by contributors drawn from a panel of doctors under the title 'Family Clinic'. This book is

a selection of 106 of these articles. The aim is not to encourage people to be 'home doctors'—indeed, there is an article warning of the dangers of self-treatment—but to produce a better understanding of illness so that unnecessary anxiety among patients and parents can be avoided, and perhaps a doctor's task can be made easier.

These articles will undoubtedly accomplish this aim for the educated reader. One must emphasise the 'educated' because the content and tone of the articles are likely to appeal to people who prefer a realistic and rational approach to an emotional and empirical one. Anyone who reads these articles will appreciate that the practice of medicine is based on general principles on which deductions are made after observation. This itself is sound health education, for the popularisation of medicine hitherto has tended to present the doctor as an infallible empiricist.

Most of the articles are concerned with child health, but there are also some which deal with adult problems, including those of the elderly. Obviously the task of selection has not been easy, but it is a pity that no article on tonsils was included. This topic was dealt with in the newspaper series and contained a deal of commonsense which should be communicated to parents as a matter of urgency. However, some of the articles present modern medical views on matters in which parents tend to cling to the medical views of two generations ago. Examples are bow legs and knock-knees, the furred tongue, thumb-sucking and aperients. It is puzzling, however, to find that the author of the article on smoking shies away from the straightforward statement that lung cancer is associated with cigarette smoking. The emphasis on personality traits– from the Yale study, of course, not from Eynseck's recent paper—may be misleading.

The article on children with cerebral palsy is a straightforward explanation of what is known today regarding causation and it is emphasised that these children need 'good medical care and understanding, not pity'. Of general interest is the article on the Bottle of Medicine. The writer is realistic and admits that specific cures form a very small proportion of the prescriptions written each year. The final article on X-rays is a sound exposition of how a policy can be arrived at which takes notice of the potential dangers of radiation and at the same time does not obstruct the use of X-rays where necessary.

This is a readable volume presented in a clear and unpretentious manner. The alphabetical arrangements and the index are helpful. Perhaps the writers have sometimes been optimistic in assuming that readers will be familiar with terms like 'stasis', but it must be emphasised again that this book is directed to the educated reader.

A. J. DALZELL-WARD

Physics and Electronics in Physical Medicine

By A. NIGHTINGALE, PH.D. *London:* G. Bell, 1959, pp. 292, 30*s*.

Medical people are proverbially allergic to physics and electronics. It therefore requires courage to write and publish a book on these subjects for consumption by doctors, even though they are specialists in physical medicine. The author of this book is to be congratulated on producing one which proves readable and intelligible to all who can lay claim to a smattering of physics and electronics, and which at the same time fully achieves its object.

The excellence of the book stems from the fact that Dr. Nightingale has taken steps to disregard matter which is of academic interest only, while stressing that which has a practical application.

It is generously illustrated with exceptionally simple and clear diagrams, while the lay-out is all that can be desired.

P. BAUWENS

NOTICES

VOCATIONAL GUIDANCE FOR THE HANDICAPPED

An International Seminar *

UNDER the auspices of the International Association for Vocational Guidance, university professors, medical consultants, psychologists, vocational guidance counsellors, employment officers, therapists and social workers from 15 different countries gathered at the Hebrew University, Jerusalem, from August 22 to September 1, 1960 to discuss vocational guidance for the handicapped. The three delegates from Great Britain were Miss M. O. Davies from the Youth Employment Service, who co-ordinates work among handicapped boys and girls in the London area; Mr. G. Brown, Senior Psychologist with the Ministry of Labour's Youth Employment Executive; and Miss M. R. Morgan, Employment Officer of the National Spastics Society.

Six main topics were covered by lectures in plenary session, by simultaneous discussion in working groups and by presentation of working groups' reports. The topics were 'Assessment of work potential as the first step in the vocational rehabilitation process'; 'The first steps in the vocational guidance of the handicapped child'; 'Vocational guidance of handicapped adults'; 'The interdependence of vocational guidance and other rehabilitation services'; 'Vocational training and placement of the handicapped'; and 'Co-ordination and integration of all services dealing with the handicapped'.

It was obvious that similar problems occur nearly everywhere, and although in some countries—for example, the U.S.A. and France—there are comprehensive counselling services, a great deal still needs to be done to co-ordinate and expand the services for handicapped people.

It was agreed at the outset of the seminar that there is a fundamental difference between the vocational guidance and training of a boy or girl with a congenital handicap and that of the man or woman who acquires a handicap later in life and needs to be rehabilitated and possibly retrained. Vocational guidance for the congenitally handicapped is a long and continuing process and prevocational training should begin as early as practicable in the child's school career. The child should learn to live with his handicap in normal surroundings and to develop positive attitudes and character traits. Activities should be directed towards developing understanding of his possibilities and adapting his aspirations to reality. Information should be given to him and his parents about occupations and trades by means of talks and visits to workshops, factories and offices.

The importance of team work in vocational counselling and the urgent need for co-ordinating and making available all the relevant data—medical, educational, psychological and social—were constantly underlined. The team should basically consist of a doctor, psychologist, vocational guidance counsellor and/or placement officer and social worker, and will include teachers, therapists and representatives from other disciplines as required. The co-ordinator of the team will vary in different settings—e.g., hospital, school or training centre—but the importance of continuity was stressed, as was the need for all members of the team to have a sound knowledge of local conditions and the general employment situation.

* A comprehensive report of the Seminar will be published later. *Editor.*

One of the team of professional workers dealing directly with the child should also be responsible for giving guidance to the parents. Much confusion is caused at present because parents are given conflicting advice by different professional workers. Parents must also be helped to recognise and face their basic attitude to the child, which may be rooted in fear, shame, guilt or other prejudices, so that they can achieve realistic acceptance of the child as a handicapped person.

A great deal more needs to be done to educate the public to accept handicapped people and help them to become fully integrated in society. Prejudices have to be combated and all outlets for training and employment must be fully explored and developed by mass propaganda, canvassing employers and placement on a case basis. It is only as these outlets are developed and known that vocational guidance becomes realistic.

M. R. Morgan

FILMS ON CEREBRAL PALSY

The 18-page catalogue of films on cerebral palsy, originally published as a supplement to *Bulletin* Vol. 1, No. 6, 1959, is still obtainable, free of charge, from the National Spastics Society at 28 Fitzroy Square, London, W.1. A list of films that have become available since the appearance of this catalogue is now being prepared and will be distributed as a supplement to *Bulletin* Vol. 2, No. 4, 1960.

Further film catalogues will be published as supplements from time to time, on Neurology, Embryology, Paediatrics, etc.

The Editor will be grateful for information on scientific films that can be used in bringing the catalogues up-to-date and making them as complete as possible.

A HUNGARIAN *ACTA PAEDIATRICA*

THIS new quarterly publication of the Hungarian Academy of Sciences is to publish paediatric papers in English, German, French or Russian, with abstracts in one of the other languages. The editorial board consists of Hungarians of international reputation—Drs. Gerlóczy, Kerpel-Fronius, Kulin, Petényi and Véghelyi; the Editor is Prof. P. Gegesi Kiss. Four issues will make a volume of about 400 pages. The subscription is £3 a year, and the sole distributors are I. R. Maxwell & Co. Ltd., of 4 & 5 Fitzroy Square, London, W.1 and 122 East 55th Street, New York 22, N.Y., U.S.A. For the sake of librarians and indexers, not to mention their journalistic colleagues, the Editors might perhaps have avoided possible confusion with the long-familiar Swedish *Acta Paediatrica* by calling their new journal '*Acta Paediatrica Hungarica*'. But whatever its name it has our best wishes.

E. Clayton-Jones

INTERNATIONAL JOURNAL OF NEUROLOGY

THIS journal, edited in Montevideo by Victor Soriano, is having a special number on Brain Vascular Diseases. The main articles include: Problems posed by cerebral ischaemic accidents in atherosclerosis, by Th. Alajouanine, F. Lhermitte and J. C. Gautier, of Paris; Haemorrhage and softening of the brain, by V. Tronconi and L. Montanari, of Ferrara, Italy; Cerebral haemorrhage, by C. D. Aring of Cincinnati; Intracranial aneurisms, by

R. J. L. Pluvinage, of Paris; The E.E.G. in cerebral ischaemic syndromes, by F. Rohmer, D. Kurtz and M. Collard, of Strasbourg; Surgical treatment of supratentorial aneurisms, by L. Pool, L. A. Mount, M. D. Yahr, J. F. Hammil and J. Ransohoff, of New York, and several others from New York. The journal's address is: Calle Buenos Aires 363, Montevideo, Uruguay.

E. Clayton-Jones

WORLD COMMISSION ON CEREBRAL PALSY

DR. GUY TARDIEU has succeeded Dr. C. D. S. Agassiz as chairman of the World Commission. In unanimously recommending Dr. Tardieu for this international post the members of the World Commission recalled his work with brain damaged children at the Raymond Poincaré Hospital in Paris and his well-known studies on the measurement of the stretch reflex in cerebral palsy. Dr. Agassiz becomes a vice-chairman of the Commission, together with Bjorn Magnussen in Denmark and Ben Epstein in South Africa; Dr. Brewster S. Miller, medical director of the United Cerebral Palsy Associations, is now Secretary of the Commission. The World Commission is composed of members from eleven different countries, and is sponsored by the International Society for Rehabilitation of the Disabled, more familiar under its old name of the International Society for the Welfare of Cripples, whose president, Mr. Hall H. Popham, announced these appointments after the Society's recent congress in New York.

E. Clayton-Jones

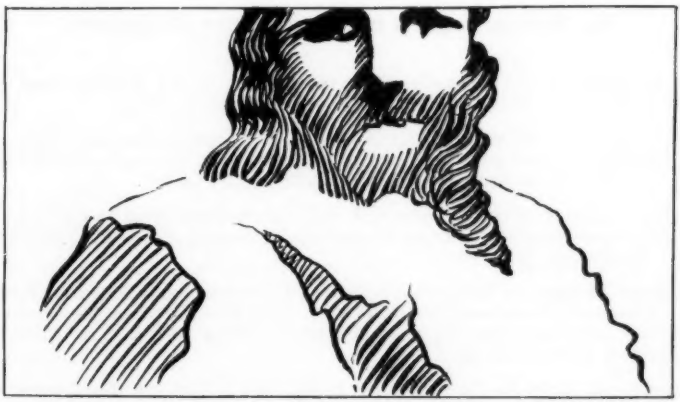


Fig. 2. M. L. JOHNSON ABERCROMBIE. 'The Hidden Man Revealed.' (*See* p. 146) (Reproduced by kind permission of the publishers, Messrs. Hutchinsons, from *The Anatomy of Judgement* by M. L. J. Abercrombie.)

PUBLICATIONS OF THE N.S.S. AND B.C.W.S.

A. National Spastics Society

BOOKLETS

The following booklets, prepared by the N.S.S. Advisory Committees, are obtainable from the Society, free of charge for single copies:

Organisation of Out-Patient and Day Clinics, Diagnostic and Therapeutic, for Cerebral Palsied Children. Medical Advisory Committee. Pp. 11.

Notes on the Assessment of Educational Needs of Children with Cerebral Palsy. Medical Advisory Committee. Pp. 7.

After School What? Notes on the training and employment of adolescent and adult spastics, with suggestions for the future, and a brief bibliography. Prepared by Miss M. R. Morgan, the Society's Employment Officer, for the Employment Committee. Pp. 9.

Lectures delivered at a Conference for those concerned with the Education and Training of Spastics, April 1955. Main speakers: Norah Gibbs, E. P. Quibell, Prof. A. V. Neale, Mrs. C. Pollack (late of Chicago), J. B. Stewart and Donna Leather. Pp. 50, 2*s.*

B. British Council for the Welfare of Spastics

BOOKLETS

These are published by and obtainable from the Council, at 13 Suffolk Street, Haymarket, London, S.W.1. The prices shown are for single copies, including postage:

The Cerebral Palsies of Childhood. H. Keith Lucas, M.Ch.Orth., F.R.C.S. A study in the recent advances in aetiology and treatment and their relation to orthopaedic surgery. Prof. Norman Capon, in a preface, commends it particularly to doctors. 1949, pp. 28, 1*s.* 9*d.*

Cerebral Palsy Today. Prof. A. A. Moncrieff, F.R.C.P. Address given at the Council's annual meeting in 1957, the main headings being Nomenclature, Sensory Findings, Education and Management, and The Future. 1957, pp. 15, 1*s.* 6*d.*

Notes on Speech Therapy, for Parents of Children with Cerebral Palsy. Shulamith Kastein, speech pathologist to Columbia Presbyterian Medical Center, New York. Based on Mrs. Kastein's '*Speech Hygiene Guidance*', and describing the methods used in training children at her Pediatric Cerebral Palsy Clinic in New York. 1952, pp. 23, 1*s.* 3*d.*

Parents' Handbook. Notes for Parents on the Home Care of Children Handicapped by Cerebral Palsy. Full of practical advice and illustrated with photographs of special equipment, many of them supplied by Miss Mayer of St. Margaret's School, Croydon. 1958, pp. 24, 2*s.* 6*d.*

Some Facts about Cerebral Palsy. Questions often asked by teachers, physiotherapists, social workers and others, with answers intended to provide introductory background information and indicate the provisions for spastics now available or likely to become so. 1959, pp. 20, 1*s.* 6*d.*

A Case of Spatial Inability in a Cerebral Palsied Child. Edith M. Caldwell. The case of Margaret, a patient in the Percy Hedley School for Spastic Children, Newcastle-upon-Tyne, is recorded in detail for teachers faced with the common problem of cerebral palsy children whose slow progress is due to lack of appreciation of space rather than faulty motor co-ordination. 1956, pp. 28, 4*s.* 6*d.*

CONFERENCE REPORTS

Addresses given at a Conference for the Staffs of Special Cerebral Palsy Schools and Units, Selwyn College, Cambridge, August, 1953. Sixteen practical talks on education and treatment. 1954, pp. 52, 2*s.* 6*d.*

Addresses given at a Three-day Conference on the Treatment of Cerebral Palsy, London, September, 1954. Speakers included W. M. Phelps, Temple Fay, M. A. Perlstein and Wylie McKissock (on hemispherectomy). Pp. 88, 3*s.* 6*d.*

Cerebral Palsy—the Present Position and Future Possibilities. Addresses at a one-day Conference, London, October, 1955. Speakers included Prof. N. B. Capon and Marjorie Wilson.

Four Addresses on Cerebral Palsy given at a Conference for Members of the Medical Profession, Birmingham, 1956. Speakers: Prof. R. S. Illingworth, Frances Braid, H. M. Cohen and A. Innes. Pp. 40, 3*s.* 6*d.*

Factors in the Assessment and Education of Children with Cerebral Palsy. Conference in Bristol, 1958. Speakers: Prof. A. V. Neale, R. M. Norman, Grace Woods, M. J. Ram, B. Bolwell, R. V. Saunders, and A. L. Smallwood. Pp. 48, 4*s.* 6*d.*

LEAFLETS

These are published by the Council at 6*d.* each.

Bringing up a Physically Handicapped Child. Dora M. Griffith.

The Parent and the Cerebral Palsied Child. Mrs. P. Robertson.

Cerebral Palsy Equipment for Children and Adults. A series of six leaflets on: (1) Chairs and tables; (2) Chair, upright; (3) Chair, tilted; (4) Table, adapted; (5) Table, adjustable; and (6) Toilet chair. With detailed instructions and working drawings.

Abstracts

In collaboration with *Abstracts of World Medicine*, published by the British Medical Association, and with the kind assistance of the Excerpta Medica Foundation, *Courrier*, and *Obstetrical and Gynecological Survey*.

Some Aetiological Problems in Mental Deficiency

J. M. BERG and B. H. KIRMAN. *British Medical Journal*, Oct. 31, 1959, **ii**, 848–852. 29 refs.

Some aetiological problems of mental deficiency are discussed with reference to the published findings of a number of workers and the authors' observations at the Fountain Hospital, London. Mental deficiency is considered under two heads—minor (high-grade) and severe (low-grade). Although psychological, educational, and social factors operate more in producing the minor mental defect, 'diluted' forms of gross pathological change can also produce minor defect. This is illustrated in cases of phenylketonuria, kernicterus, tuberose sclerosis, and the Sturge-Weber syndrome. Of a series of 44 subnormal children, a clinically diagnosed syndrome was present in 8, strong evidence of encephalopathy in 8, some sensory defect in 14, and somatic evidence of embryopathy in 2. Thus, 32 had some clinical abnormality which could account for or contribute to the mental condition. In this series of children it was also found that environment was more important than low intelligence *per se* in successful adaptation to life.

In nearly all low-grade defectives some brain abnormality is found at necropsy, but the cause can be demonstrated in only a small minority. In 200 consecutive cases of low-grade mental deficiency admitted to the Fountain Hospital there was a known cause in only 19 (9.5 per cent) and a probable cause in 8 (4 per cent). Various factors were probably or possibly implicated in 111 (55 per cent), leaving 62 cases (31 per cent) in which no causal factor was suggested. Of the 200 patients 44 were mongols, this being the largest single clinical group. Discussing the possible aetiology of mongolism the authors tentatively attribute the condition to a metabolic abnormality in the mother, becoming more manifest with increasing age, acting on a predisposed foetus. Evidence suggesting that this abnormality is connected with vitamin-A uptake is briefly mentioned. Only 32 patients in the series had no physical signs of abnormality, though evidence showed that nearly all had some gross encephalopathy.

Factors producing mental deficiency are considered under three headings: (1) those acting before conception—that is, genetic factors; (2) those acting during pregnancy—namely, maternal environment, disease, malnutrition; and (3) those acting after birth—that is, external environment. All three are likely to play a part in any one case. *E. H. Johnson*

Note: Anything which can be discovered in relation to causes or aggravating factors in mental deficiency is always important, and perhaps even more so now, in view of the implications of the new Mental Health Act. This paper gives some general guidance on possible aetiological factors, but the remarks on mongolism seem to be more hypothetical than factual, with no mention of the recently described chromosome aberrations.—*A. V. Neale*

Role of Intensive Physical and Occupational Therapy in the Treatment of Cerebral Palsy: Testing and Results

A. J. INGRAM, E. WITHERS and E. SPELTZ. *Archives of Physical Medicine and Rehabilitation*, Oct. 1959, **40**, 429–438. 8 figs., 6 refs.

At the Crippled Children's Hospital School, Memphis, Tennessee, the authors have attempted to solve the problem of how to decide which children with cerebral palsy are most likely to profit from training by devising a satisfactory objective method of testing the patients' response to treatment, based partly on the developmental schedules for normal children outlined by Gesell and partly on the findings of other authors. The test measures the child's 'motor age' and the 'social age' at 5 different ages up to 5 years and was applied after all children had been given a vigorous programme of physiotherapy and occupational therapy. The full chart of the test, which is given every 3 months, is reproduced. The authors claim that its great advantages are that it is completely objective and provides a picture of the child's progress which can be rapidly read and is not cluttered up with minutiae of treatment.

Of 60 children who were subjected to the test 40 improved with schooling and 20 did not. Of the 20 failures it was found that 12 (20 per cent) failed to respond altogether, while the other 8 made some progress and then ceased to improve; the reasons for the failures are analysed and discussed at some length. The authors conclude that this test will show when it is worth while continuing to attempt the education of a child with cerebral palsy and when such an effort will be disproportionate to the probable result. *W. Tegner*

Note: Since every child affected by some form of cerebral palsy is 'unique' in the educational and ability concept it is not altogether wise to rely on mathematical calculations of 'motor age' or 'social age', although, admittedly, some useful guidance in assessment can be derived from such data. So long as the assessments are made by real experts the suggestions made in this paper would be useful. Learning may be slow, tortuous and arduous for the affected child; teachers and physiotherapists have learnt that immense patience and a real long-term outlook is necessary. It is in any case a serious matter to write off any child as 'hopeless' or as 'case-closed'.—*A. V. Neale*

An Improved Test for Phenylketonuria

C. O. RUPE and A. H. FREE. *Clinical Chemistry*, Oct. 1959, **5**, 405–413. 9 refs.

Now that the significance and treatment of the metabolic disturbance known as phenylketonuria (phenylpyruvic oligophrenia), in which phenylpyruvic acid is excreted in the urine, are recognised, the early and efficient detection of this substance in the urine of these patients is of importance. This report describes a new strip method of testing urine for the presence of phenylpyruvic acid in which stiff strips of cellulose ('phenistix') impregnated with ferric and magnesium ions, with *cyclo*hexylsulphamic acid as a buffer, are simply dipped in the test urine. Phenylpyruvic acid produces in about half a minute a grey to blue colour, the intensity of which is proportional to its concentration; if the reaction is negative the strip remains yellow. Tests carried out by 10 experienced technicians and by 11 non-skilled persons on 1,904 specimens of normal urine to which various concentrations of phenylpyruvate had been added showed that the inexperienced as well as the experienced observers were able to recognise a level of 15 mg. of phenylpyruvate per 100 ml. of urine in 99.5 per cent of these blind tests. The tests also demonstrated the specificity of the colour reaction and that the strip method was more accurate than the liquid ferric chloride technique, which was performed at the same time. The strips are stable, and have been shown to have a 'laboratory-shelf life' of several years. Comparison with the colour chart supplied with the strips improves the specificity of the results, since it helps to obviate observer variations and a tendency to over-read the colour change. *Harry Coke*

The Relationship between Decreased 5-Hydroxyindole Metabolism and Mental Defect in Phenylketonuria

C. M. B. PARE, M. SANDLER and R. S. STACEY. *Archives of Disease in Childhood*, Oct. 1959, **34**, 422–425. 1 fig., 21 refs.

In this study reported from four London hospitals 49 patients with phenylketonuria and 32 mentally defective patients matched for weight, age and intelligence who served as controls were subjected to intelligence tests, the Terman–Merrill revision of the Stanford–Binet test, the Merrill–Palmer test, and the Vineland Social Maturity Scale being used, while the serum 5-hydroxytryptamine (5–HT) level was estimated in all the children except one control and the urinary creatinine and 5-hydroxyindoleacetic acid (5–HIAA) excretion determined in early morning specimens of urine.

The mean urinary excretion of 5–HIAA in the phenylketonuric children was 2.2 mg. per g. of creatinine whereas in the controls it was 7.2 mg. per g. of creatinine; the mean serum 5–HT levels were 71.2 and 283 mg. per ml., respectively. No significant association was found between I.Q. levels and 5–HT or 5–HIAA values. A low negative correlation was found between age and 5–HIAA excretion in both groups of patients; there was no correlation between age and serum 5–HT level, but a low positive correlation was established between 5–HT and 5–HIAA values. In some of the mental defectives the serum 5–HT concentration was as high as in the carcinoid syndrome. In discussion the authors suggest the possibility of a multiple deficiency in phenylketonuria and refer to the role of catechol amines and 2-aminobutyric acid as important substances in the physiology of the central nervous system. They are now investigating the effect of iproniazid in increasing the concentration of biologically active amines in the brain, since a decreased concentration of these substances may be responsible in some degree for the mental defect in phenylketonuria. *G. de M. Rudolf*

Emotional and Personality Development in Neuromuscular Disorders

M. L. BLUMBERG. *A.M.A. Journal of Diseases of Children*, Sept. 1959, **98**, 303–310. 11 refs.

In this paper the author (presumably a psychiatrist) presents his views about the emotional problems of children with neuromuscular diseases such as cerebral palsy, myopathies, and poliomyelitis. He expresses the view (unsupported by evidence) that the emotional problems vary according to the nature of the handicap—those of the spastic child, for instance, differing from those of the athetoid child. He discusses the attitude of those coming into contact with the child, perceptual difficulties, the lack of social experience, and the child's failure to appreciate his disabilities and their implications. *R. S. Illingworth*

Note: The author attempts to link up psychosocial and emotional patterns in children affected with spastic, myopathic or flaccid muscular disabilities. Since the background of each child is likely to be widely varied, it may well be that the emotional and reactive patterns are in fact the resultants of highly complex groups of endogenous and exogenous factors. However, doctors, teachers and therapists often seem gradually to obtain an appreciation of the child's 'inner mind' which enables them to enter all the more into his whole personality in day-to-day contacts and care, which, of course, is invaluable.—*A. V. Neale*

Teratogenic Effects of Asian Influenza

M. G. WILSON, H. L. HEINS, D. T. IMAGAWA and J. M. ADAMS. *Journal of the American Medical Association*, Oct. 10, 1959, **171**, 638–641. 12 refs.

In order to determine the effect of maternal Asian influenza on congenital anomalies in the offspring the incidence and type of deformities in newborn infants of 75 mothers with known haemagglutination-inhibition (H.I.) titres were compared with those in infants of 51 mothers without positive H.I. titres 9 months after an Asian influenza epidemic in Los Angeles County.

Only major congenital abnormalities were considered. No significant difference was observed between the two groups in the incidence of abnormal births or the incidence of congenital anomalies. Thus there does not appear to be any evidence of a teratogenic effect of infection with Asian influenza virus in the pregnant woman.

Winston Turner

Note: Scientific interest (and natural human anxiety) is constantly aware of the possible influence of viruses on human embryogenesis. Asian influenza, in its occasional global or subglobal epidemics, is obviously a disease to watch carefully. Several relatively small prospective surveys have been reported; this one is on a small scale, and any conclusions that can be drawn from it seem essentially negative. But the children must presumably be followed up for at least 5 years before one can definitely exclude late-developing aberrations of tissue growth or unsuspected effects on hearing, perception or motor capacity. The evidence so far seems to suggest that, if Asian influenza has any effects on the foetus, it is much less potent or certain than rubella or even mumps as a direct or contributory cause of congenital malformations.—*E. Clayton-Jones*

A Description and Classification of the Common Disorders of Speech in Children

T. T. S. INGRAM. *Archives of Disease in Childhood*, Oct. 1959, **34**, 444–455. 25 refs.

From experience over 3 years at the Speech Clinic of the Royal Hospital for Sick Children, Edinburgh, the author has evolved a classification of speech disorders based on the major speech defect and on associated clinical and psychological findings. It is emphasised that accurate diagnosis and classification of speech defects require a team consisting of a speech therapist, psychologist, paediatrician, and otologist. Of the 189 children studied, two-thirds of whom were under 5 years of age, only 4 had 'disorders of voicing' [they are not described in the paper]. There were 29 children with 'speech dysrhythmias', of which two types were recognised—'clutter' (3 patients) and 'involuntary temporary arrest of the flow of speech' (26). Of these 29 patients, 21 showed psychological disturbances, 9 being given psychiatric treatment. 'Dysarthric speech disorders', associated with organic disease of the central nervous system or some local lesion resulting in difficulty in articulation, were observed in 30 children. A hearing defect or mental retardation was present in 41 children who were suffering from 'secondary speech disorders'. In the largest group of 80 children 'functional speech disorders' were not associated with demonstrable disease or any abnormality of articulatory function. The group of 'mixed and unclassified disorders' contained 4 children only, including one with severe hypomandibulosis and one with cleft palate.

David Morris

Note: The function of speech is always important in the ultimate efficiency and wellbeing of a child. Hence our increasing knowledge of the cerebral and more peripheral factors concerned with the admittedly highly complex mechanisms of speech should help to distinguish cause from effect in disorders of speech. In this paper Dr. Ingram considers the classification of speech defects and rightly emphasises the importance of accurate diagnosis. Speech 'dysrhythmias' receive special mention.—*A. V. Neale*

Cerebral Palsy and Hearing Impairment. (*In Swedish.*)

B. BARR and I. KLOCKHOFF. *Nordisk Medicin*, 1959, **62**, 1512–1514, 14 refs.

It is reported from Karolinska Sjukhuset, Stockholm, that 161 physically severely handicapped children, mainly with cerebral palsy, have been examined with regard to the quantitative and qualitative presence of hearing defects. Complete audiograms were obtained by play audiometry in 150 cases. In the remaining 11 cases informal hearing tests had to be resorted to. The risk of failure to observe a

hearing impairment in these cases is emphasised, and the most common causes of misinterpretation are specified.

In all, hearing defects were found in 17 per cent. In the spastic group the incidence was 10.5 per cent, and in the athetoid group as high as 68 per cent. The localisation of these hearing defects is discussed, and the theory of a central lesion is supported by Metz's objective recruitment test, which was not positive in any of the cases.—*From Author's summary*

Note: The incidence of hearing loss in the athetoid group, given as 68 per cent, is unusually high compared with findings in this country. The sample of athetoids was comparatively small (25 of the total of 161 children examined). The total number of cerebral palsied children examined was 92 and a hearing loss was detected in 24, and this roughly indicates the same incidence of hearing loss in the cerebral palsy population at large as is found in this country.

The discussion of the site of lesion is interesting. The opinion of Fisch (1955) is quoted, who stated the reasons why the site of lesion is most probably in the cochlear nuclei (selective vulnerability of the cochlear nuclei). One author, on the basis of subjective tests of the 'recruitment of loudness' phenomenon, formed the opinion that the lesion is in the peripheral receptor. Loudness recruitment tests are supposed to differentiate between a lesion in the inner ear and in the auditory pathway. It is well known that the subjective recruitment test is often unreliable, and it is difficult to carry out in cerebral palsy subjects in any case. The interesting and new observation in the Swedish paper is the result of objective types of tests of recruitment of loudness (by measuring the alteration of acoustic impedance of the ear which occurs as a result of contraction of the stapedius muscle). The authors found that the recruitment phenomenon *does not* occur in deafness from cerebral palsy, and therefor Fisch's opinion that the lesion is not peripheral is supported by the results of these tests.—*L. Fisch*

Birth Trauma to the Vertebral Arteries

P. O. YATES. *Archives of Disease in Childhood*, Oct. 1959, **34**, 436–441. 11 figs, 31 refs.

To find out whether babies who die at about the time of birth suffer any traumatic lesions of the neck, 60 infants were examined post mortem in the Department of Pathology of the University of Manchester. They were selected at random from 213 cases of stillbirth and neonatal death occurring among 2,800 births: hydrocephalics and macerated stillbirths were excluded. The 60 infants included 16 stillborn, 8 born by breech delivery, and 3 by caesarean section; 20 were more than 2 weeks premature and 8 suffered from rhesus antibody disease. Without the details of the delivery or the rest of the post-mortem findings being known, the necks of these 60 infants were scrupulously examined histologically at various levels.

Distortion trauma of the cervical spine was present in 27 cases, the lesions being of 3 main types: (*a*) extradural and subdural haemorrhages; (*b*) haemorrhage into joint capsules; and (*c*) torn ligaments and dura: these types of trauma occurred after all methods of delivery, but were most severe after breech delivery. Direct damage to the spinal cord was found in only 2 infants, both born by breech delivery. Bruising and tearing of the spinal nerve roots were found in 9 infants. Haemorrhages in the advential coat of one or both vertebral arteries were found in 24 cases. There was thrombosis of one vertebral artery in a baby who died 12 days after birth.

The author discusses very fully the possible causative mechanisms and surveys comprehensively the implications of such lesions and the accompanying brain damage in causing such diseases as cerebral palsy and epilepsy. *David Morris*

Note: This paper contains very important observations on possible traumatic effects to the vertebral arteries during or at the time of birth. It is shown that distortion trauma of the cervical spine may be related to extradural or subdural haemo-

rrhage, haemorrhage into joint capsules, or torn ligaments and dura. These effects might occur after any form of delivery but were most severe after breech delivery. Direct damage to the substance of the spinal cord was found in two infants after breech birth. Injury to spinal nerve-roots was indicated by tearing and bruising.

The vertebral arteries were most carefully examined and the two main changes found were haemorrhages into the adventitial coats in 24 cases out of the 60 examined, and thrombosis in a vertebral artery.

The possible consequences of these traumatic lesions is properly considered and their relationships to certain forms of cerebral palsy and/or convulsive brain disorder is noted.—*A. V. Neale*

Evaluation of 'Equanil' as Adjunct to Physical Therapy for Children with Severe Cerebral Palsy

A. GELPERIN and O. PAYTON, *Physical Therapy Review*, June 1959, **39,** 383–388. 13 refs.

Meprobamate ('Equanil') was given as an adjuvant to physical rehabilitation in the management of 28 children with severe cerebral palsy at Dixon State School, Illinois. The children, aged 3 to 18 years, were suffering from athetoid or spastic cerebral palsy and their intelligence level was low. Meprobamate was given to 14 of the children and a placebo to the remainder; all 28 received physiotherapy, which included underwater exercises and active and passive movements. At the end of $2\frac{1}{2}$ months no difference was observed between the two groups, so the dosage of meprobamate was increased, this treatment being continued for a further $6\frac{1}{2}$ months. Of the 14 patients given this drug, 11 improved, although drowsiness was a problem in some of them. Improvement was noted in a similar number in the control group, suggesting that the intensive programme of treatment and the interest in the outcome of all concerned were operative factors in both groups.

The authors suggest that meprobamate may be of value in the management of certain cases of cerebral palsy. *W. Tegner*

Note: The search for some 'wonder' pharmacology for cerebral palsy is obviously worth while. However, the intricacies of the neuro-physiological and neuro-muscular components in the motor disabilities, such as athetosis, hypertonic palsy, rigidity, etc., are a long way from our real understanding. Hence trials of relaxant drugs will have to continue on a relatively empirical basis for the present. Current research is quite active in these fields. It is reasonable that clinical experimenters should try meprobamate as an adjuvant to physical therapy. However, the evidence in this paper gives barely more than a suggestion that this substance may be of some help in physical rehabilitation. It is perhaps worthy of further trials over longer periods and in different ranges of dose insufficient to produce more than minimal drowsiness.—*A. V. Neale*

The Effect of Muscle Relaxants on Cerebral and Spinal Spastic Conditions (*In German*)

W. STEINBRECHER. *Deutsche medizinische Wochenschrift*, Dec. 18, 1959, **84,** 2295–2298. 6 figs., 23 refs.

The author describes a clinical trial of a new muscular relaxant, 'Quiloflex' (a benzodioxane derivative), on patients with spastic paralysis at the City Hospital, Essen. The drug had shown strong inhibitory effects on the reflexes of the cat, and it was hoped that it would prove useful in human spastic conditions. Altogether 41 patients were treated for periods ranging from 10 days to 15 months. Electromyography shows that release of tension of a spastic muscle does not immediately follow the voluntary act of relaxation, and the time lag between these two events was measured to provide objective evidence of the degree of relaxation obtained.

Before the trial was started it was established clinically that quiloflex was contraindicated in patients with psychiatric disorders or symptoms of extrapyramidal disease. It was also found that the 13

patients in the series with cerebral spastic conditions did not benefit from the new drug, even in large doses, whereas the 28 with spinal conditions showed marked improvement. Electromyograms recorded during rest from 14 of these 28 patients, 4 of whom were treated with guaiacyl glyceryl ether (10 to 20 ml. of 5 per cent solution 3 times daily) and 10 with quiloflex (5 to 20 mg. 3 times daily) showed absence of electrical activity in the latter, whereas there was no significant change in the former. Clinical assessment correlated strongly with the patients' subjective impressions.

The results of treatment, classified according to defined criteria, were very good or good in 13 cases, definite (objective) improvement in 11, subjective improvement only in 2, and no effect in 2. It was seldom necessary to increase the dose above 20 mg. 3 times daily, the best results being obtained by starting with 5 mg. twice a day and gradually increasing it. The intramuscular route was avoided. There were no significant side-effects, and improvement in mood was thought to be due to the increase in physical well-being rather than to drug-induced euphoria. There was no disturbance of the autonomic nervous system.

[Although of very limited scope, this trial does suggest that quiloflex may prove to be a useful adjunct to the treatment of spinal spastic disease.] *M. R. Medhurst*

Note: The search for therapeutically useful muscle relaxants continues. The benzodioxane derivative, quiloflex, has here been tried in 41 patients over periods ranging from 10 days to 15 months. Given by mouth in moderate doses there were no significant side-effects and there seemed to be a favourable action on muscle tone in spinal spastic conditions. The 13 patients with cerebral spastic conditions showed no benefit. Sometimes there was a mild pseudo-euphoria, but no disturbances of the autonomic nervous system have been observed.—*A. V. Neale*

Patterns of Skeletal Development in the Hand

S. I. PYLE, R. B. REED and H. C. STUART. *Pediatrics*, Nov. 1959, **24**, 886–903. 20 figs., bibliography.

This paper is one of a series of reports on a longitudinal study of child health and development being carried out by the Harvard School of Public Health on the basis of data collected over a 26-year period. In the present part of the study a series of 66 boys and 67 girls were enrolled and radiographs of their hands taken periodically between the ages of one and 18 years. The object was 'to demonstrate the types and amounts of variability between children, as well as the consistencies manifest by individuals in the course of the skeletal maturation of the hand'. Each child was classified according to the rate of progress in skeletal age or according to the changes in rate with age compared with the norms of the group. As various patterns were identified in individual children these were grouped together and tabulated accordingly. The standard of evaluation of the radiographs varied somewhat with changes in the accepted maturity indicator scales during the period of study, but any assessment difficulty was thought to be obviated by the fact that the same person carried out the assessments throughout. The means and standard deviations were calculated for the boys and girls at each age after all the children had been assessed. The basic classification was 'moderate', 'fast', 'slow', or 'irregular'. Three screenings were carried out, and in the end there were still 6 boys and 12 girls who failed to conform to any of the final 21 possible patterns.

The results show that although normal children pass through very similar stages of skeletal growth of the hand, they do so in a variety of ways differing in the rates of progress at the different stages and age periods. Few children appear to be very irregular in progress or change frequently to any degree in their rate of progress, the commonest shift being between childhood and adolescence.

[Needless to say, this paper has to be consulted in the original for the intricate details of the study. Although it is of a highly specialised nature, it should prove of interest to all concerned with the growth and development of children.]

David Morris

Note: The normal and 'variations' within a normal range of patterns of skeletal development are under careful longitudinal study in several parts of the world: each is likely to provide valuable information of fact. In this study, at the Harvard School of Public Health, 66 boys and 67 girls have provided the radiographic material over about 18 years. Skeletal maturation of the hands was the factor studied. Some variations were noted but irregular progress is unusual, the commonest age area for shifts from an average being in the years occupied by the important growth spurt between childhood and adolescence. On the whole, healthy children pass through regular phases of radiological bone development, as are indicated in the hand.—*A. V. Neale*

Patterns of Growth in Height and Weight from Birth to Eighteen Years of Age

R. B. REED and H. C. STUART. *Pediatrics*, Nov. 1959, **24**, 904–921. 5 figs., bibliography.

The growth charts now available are based on carefully collected data and are both reliable and useful in making tentative judgments concerning the normality of a given child's growth. It is recognised, however, that the departure of any individual from the usual range of weight and height for children of his age may be due simply to a normal variation in rate of growth. In this further report from the Harvard School of Public Health the variety of patterns of growth for height and weight found among 67 boys and 67 girls are reported with the purpose of providing a better understanding of individual differences in growth from the examination of the rates of progress during selected age periods. The children were enrolled at birth and their height and weight recorded semi-annually in the early years and later annually till 18 years of age. Three arbitrary age intervals were chosen—birth to 6 years, 6 to 12 years, and 12 to 18 years. For each age interval the amount of growth which took place was calculated for each child and the children placed in order of growth increment. The rate of growth of the 17 (25 per cent) of each sex with the biggest increment was classified as 'rapid', that of the 17 with the smallest increment as 'slow', and that of the remaining 33 as 'moderate'. Each child was assigned to a pattern according to the increment category in the 3 age groups. [The original paper will need to be consulted for the details of the groupings.] The most frequent pattern groups for both boys and girls were rapid or moderate for the early period, moderate for the middle period, and moderate or slow for the late period; this accounted for 75 per cent of the children of each sex. The rate of growth during early childhood appeared to be associated with, but not specifically predictive of, the size at maturity and timing of the adolescent growth spurt. Those with rapid growth before 6 years tended to have large mature size and an early adolescent growth spurt.

David Morris

Note: This is another of the publications from the Harvard School of Public Health on patterns of growth from birth to 18 years. Chronological and velocity data are given, and, as usual, these are interesting and serve as yet another contribution to the general biological factors concerned and indicate particular trends in individual children. There are some useful observations on the possible predictive aspects of growth before the age of 6 years and of an early adolescent growth spurt.—*A. V. Neale*

Intrathecal Phenol to Relieve Spasticity in Paraplegia

P. W. NATHAN. *Lancet*, Dec. 19, 1959, **ii**, 1099–1102. 3 figs., 2 refs.

Experience in the relief of spastic para-

plegia and its associated muscle spasms by the intrathecal injection of a solution of phenol, which is designed permanently to destroy the anterior nerve roots, is reported in this paper from the National Hospital, Queen Square, London. A solution of 20 per cent phenol in glycerol or 25 per cent in 'Myodil' (ethyl iodophenylundecanoate) is used, the amount of solution injected depending on the number of roots to be destroyed. The procedure is painless and takes half an hour. No untoward reactions to the injection, such as meningitis or phenol poisoning, were observed and there were only insignificant changes in the protein content of the cerebrospinal fluid. If the object of treatment is to destroy the 5th lumbar and 1st and 2nd sacral nerve roots, there is considerable danger of paralysing the sphincters also. In addition, there is some loss of sensation in the dermatomes of the treated roots with liability to injury of the analgesic skin and consequent development of bed-sores. After the injection attention must be paid to the sphincters.

In 25 patients with various neurological diseases who were treated there was a satisfactory response to treatment, with relief of painful muscle spasms and conversion of spastic and contracted limbs into flaccid limbs. This greatly facilitated nursing, and the patients were able to get up into easy chairs and to go out in wheel-chairs. In some cases surprising voluntary power was unmasked by the diminished spasticity. A further injection within a year because of the return of spasticity was necessary in only one of the 25 cases.

R. Wyburn-Mason

Note: In the management of some forms of spastic paraplegia associated with severe painful muscle spasms, intrathecal injections of 20 per cent phenol in glycerol or 25 per cent in myodil has relieved the spasticity, and in come cases there has been a surprising unmasking of voluntary power by the diminution in spasticity.—*A. V. Neale*

Intrathecal Phenol in the Treatment of Reflex Spasms and Spasticity

R. E. KELLY and P. C. GAUTIER-SMITH. *Lancet*, Dec. 19, 1959, **ii**, 1102–1105. 2 figs., 13 refs.

Intrathecal injections of phenol were given in the treatment of 32 cases of paraplegia with reflex spasms at St. Thomas's Hospital, London. The solutions used were 1.5 to 3 ml. of 5 to 20 per cent phenol in 'Myodil' or 1 to 2 ml. of 5 per cent phenol in glycerol, the injection site being the L2 to L3 interspace. Altogether 36 injections of each solution were given. Complications included moderately severe headache lasting 24 hours in 2 patients, bladder disturbance in 6 (long-lasting in 2), and permanent numbness of the legs in 9. A severe sensitivity reaction to myodil was observed in 2 patients, both of whom had had injections of myodil before.

Of the 32 patients, 11 were much improved and 15 were improved as regards spasticity; there was no change in 5 and one became worse. In 16 cases severe spasms were abolished and in 7 others they were much reduced and became painless. There were 2 deaths in the series, both from carcinoma, the deaths occurring too soon after the injections to permit assessment of the results. Over a follow-up period of 10 months, 4 patients had a recurrence of spasms which responded to reinjection. Reduction of spasticity revealed a surprising amount of voluntary power and gave greater comfort and improved sleep, some patients being able to sit out of bed in chairs or to stand again. In one case there was remarkable psychological improvement and in 2 troublesome bed-sores healed.

R. Wyburn-Mason

Note: This is a similar report to that of P. W. Nathan, and again intrathecal injections of 5–20 per cent phenol have had some remarkably favourable effects on reflex spasms and spasticity. There was definite improvement in 26 out of 32 patients. The partial or considerable relief

of spasticity obtained enabled a few patients to sit out of bed in a chair or even to stand again. Psychological improvement may be an additive gain.

A further paper on this subject has since been published in the *Lancet* (1960, **ii**, 31), by L. A. Liversedge and R. M. Maher, who have treated 32 patients with intrathecal injections of 2 per cent or 5 per cent phenol in glycerin. Most of their cases (21) had multiple sclerosis, but there were 3 with parkinsonism, 2 with syringomyelia, 3 with hemiplegia of vascular origin, 2 with a spastic paraplegia, probably from disseminated sclerosis, and 1 with amyotrophic lateral sclerosis. All but 6, who were severely spastic in the lower limbs, were confined to their bed or a chair; and the only one who did not have severe painful extensor or flexor spasms was the patient with syringomyelia, whose leg was in constant though painless extensor spasm. The treatment was not an unqualified success, but it appeared to be 'of circumscribed value' in the relief of painful spasms in spastic cases. Careful positioning of the patient is essential, and there is always the possibility of causing retention of urine. One should be careful about giving phenol injections to patients who are still ambulant. The treatment is especially suitable for immobile patients suffering severely from painful flexor and/or extensor spasms.

In a letter to the *Lancet* (1960, **ii**, 265) Ivan T. Draper of Edinburgh records one disturbing experience with intrathecal phenol, out of a series of 15 injections. They gave a man of 64 an intrathecal injection of 1.5 ml. of 2 per cent phenol in glycerin, with immediate relief of the spasticity, caused by disseminated sclerosis, from which he had suffered for 20 years. Half an hour later he had agonisingly violent flexor spasms, severe enough to make this stoical man cry aloud. The pain was relieved with intramuscular pethidine, and apart from a temporary dull ache there was no recurrence. He was free from painful spasms for the subsequent 3 months.

E. Clayton-Jones

Psychological Effects of Hospitalization in Infancy

H. R. SCHAFFER and W. M. CALLENDER. *Pediatrics*, Oct. 1959, **24**, 528–539. 9 refs.

The psychological effects in infancy of a stay in hospital were studied in 76 infants, aged 3 to 51 weeks, admitted to the Royal Hospital for Sick Children, Glasgow. Infants suffering from marasmus, gross deformities, or brain injury, and those who were fully breast-fed were excluded. The duration of stay varied from 4 to 49 days, average 15.4 days.

It was observed that infants under 7 months old differed considerably from those over that age in their response to separation from the mother. With a few exceptions the children under 7 months accepted separation without observable disturbances, displayed normal mobility when in the hospital, responded normally to observers, and showed an interesting but brief pre-occupation with their surroundings on their return home. In contrast, infants over 7 months of age cried vigorously when separated from the mother, displayed over-activity or under-activity (the withdrawal syndrome), sucked their thumbs, suffered from feeding disturbances, were entirely negative in their response to the staff, and on return home showed insecurity in the form of over-dependence on the mother. *R. S. Illingworth*

Note: Evidence is accumulating that it is always unnatural for an infant, whether in sickness or in health, to be removed from his mother. Common sense and psychology govern this notion. To suggest that an infant under 7 months finds separation innoxious, whereas after that age it is more liable to be noxious is perhaps a wide generality. The fact that the older infant appears more affected does not prove that the younger one is less inwardly deprived. It is doubtful if anyone can yet say what occult influences such deprivation may have on an infant under six months of age, even though the separation lasts only 4–49 days. We should keep an open (and wholly receptive) mind on this difficult

matter, perhaps acting on the idea that the less the infant under one year of age is separated the better for the infant, and usually for the mother also.—*A. V. Neale*

Psychological Learning Disorders in Children

H. R. MYKELBUST and B. BOSHES, *Archives of Pediatrics*, 1960, **77**, 247–256.

The term 'psychoneurological disorders' is used to cover all aberrations of behaviour which have a neurological basis, irrespective of age of onset and aetiology. These disorders may be present in people who are otherwise of average or above average intelligence and there may be minimal neurological signs.

Five cases are reported of children with respectively expressive aphasia, receptive aphasia, dyslexia, dyscalculia and disorientation in space and time. The child with receptive aphasia had a history of Rh factor with jaundice, and there were indications of pre- or peri-natal anoxia in the other cases. EEG was abnormal, usually mildly so. The IQ was within normal range. Educational therapy is reported as having good effects. The need for early specific diagnosis is stressed. This is part of an ambitious research project which has now been running for 3 years. Children with learning difficulties are given an examination taking 8–10 hours, and the neurological, electro-encephalographic and behavioural findings are coded and treated statistically through an electronic computer. Preliminary findings are that in a public-school population more than 5 per cent of children may have psychoneurological learning difficulties. The disorders are five times more common in males than in females. Children with problems of revisualisation tend to have disturbances in the occipito-parietal area; those with problems of re-auditorisation, in the temporal-parietal area. The neurologist in future will be able to predict learning disorders. Certain psychometric procedures indicate the area of brain involved. A scientifically orientated educational language therapy can be planned according to the specific type of psychoneurological disorder. *M. L. J. Abercrombie*

Note: It is encouraging to hear of this long term collaborative and therapy-orientated study of specific learning difficulties. It will be interesting to see if the behavioural improvements which follow therapy are accompanied by favourable changes in the EEG signs. If the neurologist can learn to detect proneness to learning difficulties before the children are old enough to demonstrate them, it may be possible with understanding management to avoid the development of some of the emotional difficulties that tend to result from learning disorders.—

M. L. J. Abercrombie

Association between Maternal Disease during Pregnancy and Myopia in the Child

P. A. GARDINER and G. JAMES. *British Journal of Ophthalmology*, 1960, **44**, 172–178.

Some workers have noted an association between congenital myopia and prematurity. The authors have studied two series of consecutive cases of congenital myopia (more than —4 D) from the point of view of maternal disease during pregnancy, and compared the findings with those in a control group of mothers and children suffering from visual disorders other than myopia. Toxaemia (hypertension, pre-eclampsia, or renal disease) occurred in 50 per cent of the mothers of the myopic children, and a further 25 per cent had other well-defined disease. Only 13 per cent of the mothers in the control group had had toxaemia and a further 12 per cent other diseases, usually of a trivial nature. Prematurity was not a feature of the myopic group. It is considered that congenital myopia may be analogous to acquired myopia, since there is evidence that it occurs in a child whose general growth is normal but whose nutrition is abnormal. *E. S. Perkins*

Note. This study of 38 cases and a control group seems strongly to support the view that maternal disease in pregnancy is sometimes a factor in the causation of congenital myopia. In an annotation on this paper, the *Lancet* (July 23, 1960, p. 194) urged the need for a long-term follow-up study of congenital myopia in full-term and premature infants, diagnosed in the first two years of life and excluding cases with a family history of myopia. Such a prospective survey would make it possible to correlate defects in the child with the clinical history of pregnancy, delivery and postnatal development.—

E. Clayton-Jones

MAIN CONTENTS OF BACK NUMBERS

Vol. 1, No. 1: 1958

William John Little: A Brief Biography — W. J. Bishop
On the Influence of Abnormal Parturition, Difficult Labours, Premature Births, etc. (Reprinted from the *Transactions of the Obstetrical Society of London*, 1861–62) — W. J. Little

Vol. 1, No. 2: 1958

Spasticity and the Intellect — The late H. Charles Cameron
'The Author': Personal Note on Dr. Cameron — T. B. Layton
Early Diagnosis of Cerebral Palsy — R. S. Illingworth
Some Special Learning Difficulties — Jessie M. Williams

EDITORIALS

Definitions and Classifications — Ronald Mac Keith
Peripheral Surgery — Lipmann Kessel
Was Little Right? — A. V. Neale

REPORT

Value of Films in Cerebral Palsy Centres — A Conference at Guy's Hospital

Vol. 1, No. 3: 1958

Muscle Sense Organs and Disorders of Movement — Geoffrey Rushworth
The Problems and Needs of Parents of Cerebral Palsied Children — M. Kathleen Sykes

EDITORIALS

Electroencephalography in Cerebral Palsy — G. Pampiglione

BOOK REVIEWS

Cerebral Palsy in Childhood — Grace Woods
Primer of Cerebral Palsy — J. D. Russ, H. R. Soboloff

Vol. 1, No. 4: 1958

Oxford Study Group on Child Neurology and Cerebral Palsy — Mary Capes
Discussion on Body Image
What the Participants Said (with group photograph)
Disturbance of the Body Image — W. Ritchie Russell
The Development of the Body Image — Grace E. Woods
Recognition and Treatment of Disturbances of the Body Image — Stella Albitreccia
Susceptibility of Brain Cells — T. E. Oppé
Charts of Heights and Weights of Children — Paul E. Polani

Vol. 1, No. 5: 1959

Present State of Child Neurology — C. Koupernik
Characteristics of Spasticity in Children — Ronald Mac Keith
Muscle Tone and Posture in Infancy — T. T. S. Ingram
Educational Disabilities of Cerebral Palsy Children — R. V. Saunders
Neurological Examination of the Newborn according to André-Thomas — Paul E. Polani

EDITORIALS

Neurology of the Newborn — Ronald Mac Keith
Definition of Cerebral Palsy — R. Mac Keith, I. C. K. Mackenzie, P. E. Polani
Discussing Terminology — M. L. Johnson Abercrombie

LITTLE CLUB MEMORANDUM ON TERMINOLOGY

Comments on the Little Club Memorandum — Paul E. Polani, R. S. Illingworth, Adelaide Grisoni-Colli, Sven Brandt, I. C. K. Mackenzie
Working Party Discussion — T. T. S. Ingram
Nomenclature and Classification: General Report — Paul E. Polani
General Discussion on Memorandum — W. Ritchie Russell, E. A. Carmichael, Ronald Mac Keith, Paul E. Polani, G. A. Pollock

BOOK REVIEW

Mental Deficiency: The Changing Outlook — Ann M. and A. D. B. Clarke

Vol. 1, No. 6: 1959

Reticular Formation Revisited — I. Calma
Problem Solving and Mental Defect — N. O'Connor
Teaching Imbeciles Industrial Skills — Ann M. Clarke
Movement of Films between Countries — Frank Bamping
Cortical Lesions in Cerebral Palsy — Leonard Crome
Results of Cerebral Surgery for Motor Disorder — John A. V. Bates
Note on the Clasp-Knife Phenomenon — Ronald Mac Keith
Hypertonus and Physical Therapy — J. H. Crosland
Discussion on Dr. Crosland's paper — William Dunham

BOOK REVIEW

L'Equilibre Mental — C. Koupernik

SUPPLEMENT

Scientific Film Association Catalogue of Films on Cerebral Palsy

Vol. 1, No. 7: 1959

Experimental Observations on Hypertonus:

1. *Chairman's Introduction* — G. G. Phillips
2. *Hypertonus and the Gamma Motoneurones* — P. B. C. Matthews
3. *Functional Disorder in Hypertonia* — G. Rushworth

Clinical Recognition and Differentiation of Hypertonus:

1. *Orthopaedic Surgeon's Outlook* — G. A. Pollock
2. *Polymyographic Studies* — G. Pampiglione
3. *Recording Dynamometer in the Assessment of Motor Function* — R. G. Willison
4. *Stretch Reflex in Man* — Guy Tardieu, P. Rondot, J. C. Dalloz, J. Mensch, C. Monfraix

Rehabilitation Centres — Edward Samuel
Late Reading in Children — William Calvin Barger
Effects of Abnormal Brain Development on Function — Paul E. Polani
Medical Aspects of a Comprehensive Survey of Cerebral Palsy — R. G. Mitchell
Habilitation of Adult Spastics — C. D. S. Agassiz

BOOK REVIEWS

The School Health Service — S. and Vera Leff
Adolescence to Maturity — V. C. Chamberlain
Mental Retardation — Hans Mautner

Vol. 1, No. 8: 1959

André-Thomas—A brief biography, with portrait — C. Koupernik
Integration in the Infant — André-Thomas
The Neuropathology of Cerebral Palsy and its Importance in Treatment and Diagnosis — K. Bobath
The Neurological Department of the District Children's Hospital in Brno — C. J. van der Gaag

BOOK REVIEWS

The Borderline of Embryology and Pathology — R. A. Willis
Recent Advances in Paediatrics — Edited by Douglas Gairdner
The History and Philosophy of Knowledge of the Brain and its Functions — Edited by F. N. L. Poynter
The Cranial Nerves — Alf Brodal
The Birth of Normal Babies — Leon P. Strean

Vol. 2, No. 1: 1960

Hughlings Jackson—A brief biography, with portrait — W. J. Bishop
Early Days of the National Hospital, Queen Square — Macdonald Critchley
The Contribution of Hughlings Jackson to Neurology — Macdonald Critchley
The 'Floppy' Infant — John N. Walton
Muscle Biopsy in the Diagnosis of the 'Floppy' Infant — A. L. Woolf
Factors Rendering the Cerebral Palsied Child Capable or Incapable of Benefiting from Formal Education — H. B. Davies

EDITORIALS

The Writings of Hughlings Jackson — R. G. Mitchell
Respiratory Distress Syndrome of the Newborn — T. E. Oppé
New Light on the Plantar Reflex — John A. V. Bates
An Operation for Paralytic Hip Contracture — J. S. Batchelor

REPORT

Sensory, Auditory and Speech Defects in Cerebral Palsy. Meeting of Heads and Therapists of N.S.S. Schools — J. P. M. Tizard, L. Fisch, D. E. Back

BOOK REVIEWS

The Anatomy of Judgement — M. L. Johnson Abercrombie
Recent Neurological Research — Edited by A. Biemond
Modern Ideas of Physical Education — M. W. Randall
The Principles of Exercise Therapy — M. Dena Gardiner
Psychosomatics — Edited by J. Booij
Lectures on Epilepsy — Edited by Lorentz de Haas

Vol. 2, No. 2: 1960

Obstetric Viewpoint on Cerebral Palsy — James Walker
Obstetrical Features related to Cerebral Palsy — G. Gordon Lennon
Comments on the papers of Professor Walker and Professor Lennon:
Comment No. 1 — A. V. Neale
Comment No. 2 — Benjamin Pasamanick
Electroencephalography in Cerebral Palsy — Arne Lundervold
A Cerebral Palsy Centre in the Tropics — C. Elaine Field
A Problem, and some Replies from: N. S. Alcock, Marcel d'Avignon, D. P. Beaman, G. A. Pollock, Guy Tardieu, Grace Woods; with a Postscript by Dr. Field.
The New Mental Health Act — Robina S. Addis
Research backed by the National Spastics Society — Alison D. MacDonald

REPORT

Developmental Disorders of the Central Nervous System. A meeting at Marienbad, Czechoslovakia. — Ivan Lesny

EDITORIALS

Abdominal Decompression in Labour — T. L. T. Lewis
Conservatism in the Management of Foetal Distress — S. D. Perchard
Classification of Mental Retardation — M. J. Craft
Mental Retardation Research Methods — M. J. Craft
Three Papers on Speech Disorders — Leonard Willmore
Psychological Services for the Cerebral Palsied: Discussion at Miami — J. M. Williams
Phenylketonuria — J. N. Cummings
Views from Australia — Thomas Oppé

BOOK REVIEWS

Speech and Brain Mechanisms — Wilder Penfield, Lamar Roberts
Outline of Human Genetics — L. S. Penrose
Speech and the Development of Mental Processes in the Child — A. R. Luria, F. I. Yudovich
The Natural History of Cerebral Palsy — Bronson Crothers, R. S. Paine
Advice to the Expectant Mother — F. J. Browne, J. C. McLure Browne
Before and After Childbirth — Jane Madders
Having a Baby — J. F. Robinson

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A DOCTOR is wanted to start a rehabilitation centre for 'spastics' in Malaga. Details from Dr. José Blasco Llopis. Siete Revueltas, 4-20. Malaga. Spain.

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Back

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